Middle East Immunology Summit of the Gulf Cooperation Council Association of Immunology and Rheumatology (GCC AIR), 2023 Dubai, United Arab Emirate. Scientific abstracts and case reports

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Introduction

The Gulf Cooperation Council (GCC) Association of Immunology and Rheumatology (GCC-AIR) held its first Middle East Immunology summit (MIS) in Dubai in the period between 3rd-4th November 2023. The MIS program focused on basic and transitional science of inflammatory arthritis, connective tissue diseases, vasculitis and autoinflammatory disorder. The scientific committee reviewed and accepted abstract from across the region. The abstracts were scored based on the merit of the following 5 criteria; (i) title, introduction, and abstract, (ii) case description, (iii) discussion, (iv) novelty and learning points, and (v) overall score. The scores were grade from 1-5 where 1 reflects very poor quality, 2: poor quality, 3: average quality, 4: good quality and 5 reflects excellent quality. Each abstract was reviewed by 3 reviewers and total score of all the reviewer was considered. Abstracts with low scores or generally poor quality according to reviewers were rejected. After peer review process, the scientific committee accepted 35 abstracts. In this review, the abstracts were presented respectively in order of their scores starting from highest to lowest.

Meeting Highlights

Category: Case reports/series

MIS2023-A-1039
Takayasu's Arteritis Presenting as Subclavian Steal Syndrome

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HIGHLIGHTS FROM MIDDLE EAST IMMUNOLOGY SUMMIT OF THE GCC AIR

Introduction: Takayasu arteritis is a chronic idiopathic granulomatous large-vessel vasculitis that affects the aorta, its main branches, and pulmonary arteries. It typically affects young women and is often not diagnosed until late in the disease because of its rarity, often indolent course, nonspecific early symptoms, and lack of specific diagnostic markers. The subclavian steal syndrome occurs due to the reversal of flow in the vertebral artery toward the subclavian artery (during periods of increased demand) because of stenosis/occlusion of the proximal subclavian artery.

Case report: A 40-year-old man was admitted with vertigo and intermittent pain over the left upper limb, which worsened with exertion. He denied history of fever, weight loss, drug abuse, and addictions. He had no other relevant medical illness in the past. Peripheral pulsations were feeble in the left upper limb and bilateral lower limbs (popliteal, posterior tibial and dorsalis pedis). Blood pressure was 140/80 mm of Hg in the right upper limb and 90/60 mm of Hg in the left upper limb. The rest of the systemic examination was unremarkable.

Biochemical parameters including electrolytes, renal function tests, liver function tests, and blood counts were normal. Erythrocyte sedimentation rate was 30 mm/hr1 and C reactive protein (CRP) was high (11.68 mg/L). Electrocardiogram, chest x-ray and trans-thoracic echocardiogram were normal. Arterial Doppler showed reversal of flow in the left vertebral artery, absent flow in the left common carotid, bilateral internal carotid, and bilateral superficial femoral artery occlusion. HIV, venereal disease research laboratory test, hepatitis B and hepatitis C serology were negative. Autoantibody profiles including antinuclear antibody, rheumatoid factor, c-antineutrophil cytoplasmic antibody, p-antineutrophil cytoplasmic antibody, and anti-phospholipid antibodies were all negative. Serum complement levels were normal. Digital subtraction angiography showed total occlusion of the right internal carotid artery, left common carotid, and left subclavian artery. On injecting dye to the vertebral artery extensive collaterals from posterior circulation were visible and the left upper limb was receiving blood supply by subclavian steal.

The patient was diagnosed with subclavian steal syndrome secondary to Takayasu's arteritis. He was started on corticosteroids (which was tapered down later), and antiplatelets. He was referred for further management.

Conclusion: We present a case of takayasu arteritis which presented with subclavian steal which is extremely unusual. The case reminds the readers to be aware of such rare presentations of Takayasu's arteritis which help them to early recognise the disease for prompt management.

MIS2023-A-1041
A Confusion with A Positive Antinuclear Antibody Test

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Introduction: Diagnosis of neuropsychiatric manifestations of systemic lupus erythematosus (SLE) remains challenging due to the nonspecific nature of the manifestations and inability of diagnostic modalities to differentiate neuropsychiatric lupus from other illnesses with similar presentations. We present a patient with acute encephalitis and a positive antinuclear antibody test then we raise the question of whether it was caused by SLE.

Case report: A 33-year-old Brazilian female was admitted with 4-day history of sleep disturbance, insomnia, anxiety, fatigue, and headache, a one-day history of confusion, behavioral changes, and a witnessed generalized tonic-clonic seizure. On examination, she was febrile (38.5°C) and tachycardiac (118 sinus rhythm). Glasgow Coma Scale was

14/15 due to confusion. Neurological examination was otherwise normal.

Initial investigations did not lead to a certain diagnosis. However, the possibility of neuropsychiatric lupus was raised after antinuclear antibody (ANA) test requested by the neurologist came back positive with a titer of 1:1600 speckled pattern (normal up to 1:200). Accordingly, the rheumatologist was consulted for the possibility of neuropsychiatric lupus. A final decision was made after a thorough discussion to manage the patient as a case of SLE with neuropsychiatric lupus. However, subsequent investigations changed the diagnosis to anti-NMDA receptor encephalitis secondary to ovarian teratoma.

Conclusion: This case highlights the importance of maintaining a broad differential for patients with encephalitis in possible cases of neuropsychiatric lupus. It also illustrates the importance of interpreting a positive ANA in its clinical context.

MIS2023-A-1018
Sacroiliitis During Isotretinoin Treatment, A Case Report

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Introduction: Isotretinoin, a potent retinoid medication widely used in the treatment of severe acne. There are many side effects associated with isotretinoin use ranging from well-documented dermatological side effects like dry lips and cheilitis to less common systemic manifestations.

Sacroiliitis can arise from rheumatic and nonrheumatic causes such as spondyloarthropathies, osteoarthritis, pregnancy, and trauma. However, there has been limited attention in the literature to the rare occurrence of sacroiliitis as an adverse effect of isotretinoin. The precise mechanisms remain poorly understood, but the emergence of several case reports suggests a potential link in immunomodulation through the alteration of cytokine balance and degradation of synovial membranes in joints.

Case report: A 19-year-old male was started on a 20 mg daily isotretinoin therapy for his acne vulgaris, which appeared on the face and upper trunk. After one month, he began to suffer gradually from pain in his low back and hip which became severe impairing his mobility. Associated symptoms include alternating buttocks pain, nocturnal pain, and early morning stiffness lasting less than 30 minutes. He has no personal or family history of rheumatological diseases. On examination; his vital signs were stable and hip movements were painful, with limitations in internal and external rotation and a positive FABER test. Lumbar flexion was restricted and painful. Schober's test was measured as 2 cm.

Medical investigation showed an elevation of erythrocyte sedimentation rate (ESR): 66 mm/h, and C- reactive protein (CRP): 81 mg/l. HLA B27 was negative. X-ray radiography of sacroiliac joint was normal, but the Magnetic resonance imaging examination showed the presence of bilateral sacroiliitis.

Following the exclusion of infectious etiologies and discontinuation of isotretinoin, along with the administration of NSAIDs, the pain in the lumbar and hip areas was alleviated. Subsequently, the inflammatory markers; ESR and CRP decreased to 12 mm/h and 9.6 mg/dl, respectively. He was lost to follow-up due to insurance-related problems and presented a year later unable to stand and walk for 3 days. His updated MRI revealed active on top of chronic bilateral sacroiliitis, accompanied by elevated inflammatory markers (ESR, 21 mm/h CRP, 17 mg/l). He was once again prescribed NSAIDs with a good response.

Our management of isotretinoin-induced sacroiliitis follows guidelines adapted from the ASAS-EULAR algorithm for axial spondyloarthritis. The algorithm entails an initial treatment with NSAIDs if no contraindications and to be titrated up to the maximum effective dose before considering biological/targeted synthetic disease-modifying antirheumatic drugs.

Conclusion: We describe a rare case presented with a clinical picture compatible with sacroiliitis potentially induced by isotretinoin in a young male. This case highlights the rare but noteworthy association between isotretinoin therapy and sacroiliitis. In patients presenting with unexplained musculoskeletal symptoms during isotretinoin treatment, clinicians should raise suspicions on the adverse effects of medication. Furthermore, this case report contributes to the growing body of knowledge regarding rare adverse effects of isotretinoin, in an attempt to allow further research to elucidate the immunomodulatory pathways involved.

MIS2023-A-1020

Brachial Artery Thrombosis after Upadacitinib Treatment in Psoriatic Arthritis Patient

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Introduction: Since the oral surveillance data, there has been some concern about major adverse cardiac events and thromboembolic adverse event (MACE) in high risk patient. There has been low attention on arterial thrombosis although oral surveillance data suggest that the interquartile rate is 0.4% (95% confidence interval 0.3 to 0.5) for arterial thromboembolism.

Case report: We report a 90-year-old female with high body mass index (42) but no other known history of venous thromboembolism (VTE) or MACE. The patient was wheelchair bound due to known lumber radiculopathy and complicated lumber laminectomy surgery. She has no other background comorbidities such as hypertension or diabetes. The patient was diagnosed as a psoriatic arthritis based on strong family history of psoriasis and psoriatic arthritis (daughter). She has negative CCP and RF but positive ANA 1:1000 and low positive smRNP (11 [0-100]), ESR (30-42mm/hr) and CRP of 11 mg/ L. She had a high DAS- 28-CRP of 5.2. Patient has and have high urate (461 umol/L) and is on allopurinol 100 mg. Patient originally presented with severe subacromial bursitis in the right side for which she had been diagnosed as polymyalgia rheumatica although she failed systemic steroid including prednisolone (up to 20 mg) and Intramuscular methylprednisolone injections. She continued to have huge subacromial collection of fluid in the subacromial bursa reach 300 ml on aspiration and failed all aspirations and corticosteroid injections and required monthly arthrocentesis. Synovial aspirate was negative for crystals and cultures were negative. After 12 months, disease progressed to cause synovitis in wrists and multiple metacarpophalangeal joints. She failed hydroxychloroquine 200 mg daily. In the final quarter of 2020 patient was prescribed Updadacitinib with the dose of 15 mg daily (before the oral surveillance data become widely accepted) as added to the ongoing treatment with hydroxychloroquine 200 mg daily and her prednisolone was escalated from long- term 5 mg to 20 mg daily to treat the ongoing flare of disease. As part of the screening the patient was found to have hepatitis B core antibodies positive but with negative hepatitis B virus polymerase chain reaction. She was given Entecavir 0.5 mg as an antiviral prophylaxis. Three weeks after starting Updadacitinib patient was admitted with severe right wrist and hand pain and on examination right hand was cold with absent radial pulse.

Arterial doppler ultrasonography of the left upper limp where all examined arterial segments reveal diffuse atherosclerotic changes in the form of mild diffuse intimal medial thickening. There was sudden interruption of the

arterial flow is noted in the distal left brachial artery, just proximal to the cubital fossa, where a thrombus/embolus is seen completely occluding its lumen. No arterial flow is seen in the distal left brachial artery, to the radial and ulnar arteries at the level of the wrist left upper limb angiography showed brachial artery filling defect and occlusion at the elbow joint.

An urgent intervention by interventional radiologist was taken where EKOS catheter was introduced and 5 mg Alteplase was administered directly as a bolus. After 24 hours patient had check angiography of left brachial artery showed good improvement in the flow in the radial and ulnar arteries but filling defect was noted in the distal radial artery with abrupt cutoff and another filling defect was also noted in the origin of the left ulnar artery causing partial obstruction. Subsequently 2 mg alteplase and 5000 IU heparin was administered intraarterially. Aspiration thrombectomy of the filling defect in the ulnar artery was performed with 6 F guiding sheath. White clot was noted in the aspirate. Patient continued to do well after that and recovered well with return in blood flow in the left arm. Upadacitinib was stopped after that. As a follow up investigation including protein C and S, B2 glycoprotein and anticardiolipin immunoglobulin G and IM and lupus anticoagulant were normal.

Conclusion: Arterial thrombosis in a case of psoriatic arthritis with the risk factors of old age, obesity, low mobility and corticosteroid use. Although no direct causation could be established, but caution might be considered in the presence of the risk factors of low mobility and obesity in elderly with Janus kinase inhibitors.

MIS2023-A-1036

A Confusion with a Positive Antinuclear Antibody Test

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Introduction: The C3 glomerulopathies refer to a set of uncommon kidney disorders marked by irregular complement activity. This leads to significant deposition of C3 complement in kidney histology. The primary subgroups within C3 glomerulopathy, namely dense deposit disease (DDD) and C3 glomerulonephritis (C3GN), exhibit similar clinical and pathological traits, indicating a potential continuum of the disease. We are reporting a case of biopsy proven C3GN presented to our institution. Our objective is to enhance awareness of this rare condition and shedding light on some complex pathophysiologic and genetic aspects of the disease and importance of genetic testing in such cases.

Case report: A 21-year-old female, was originally admitted at age of 12 years with proteinuria and history of presumed post streptococcal glomerulonephritis in 2012 which was treated with Angiotensin-converting enzyme inhibitors (ACEI) enalapril and was followed up by paediatric nephrology team. Her proteinuria persisted and she was advised for a renal biopsy for further evaluation which was declined. She stopped enalapril after about a year and was lost to follow up until she presented 9 years later with hypertensive urgency and severe renal impairment. Her laboratory test results included creatinine 355 micromol/L, protein/creatinine ratio 4.5 g/g creatinine (nephrotic range proteinuria). Initial urine analysis showed some microscopic haematuria (RBC 87cells/microL), no casts or active sediments seen. She tested negative for viral/ hepatitis

Conclusion: We reported a patient with C3 glomerulopathies which is often not diagnosed promptly, potentially resulting in adverse health outcomes. It is a disease to look for whenever there is kidney disease with unknown etiology. More data is needed on the topic given the rarity of its occurrence and more cases should be published to emphasize the significance of early diagnosis and management.

HIGHLIGHTS FROM MIDDLE EAST IMMUNOLOGY SUMMIT OF THE GCC AIR

MIS2023-A-1025

Clinical Characteristics and Outcomes of Chronic Nonbacterial Osteomyelitis in Children: A Single, Tertiary Centre Case Series in Abu Dhabi, UAE

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Introduction: Chronic nonbacterial osteomyelitis(CNO) is a primary autoinflammatory bone disease that presents more frequently in children and is characterized by inflammatory bone lesions in the absence of an infectious etiology. The aim of this case series is to evaluate the demographic, clinical, laboratory, imaging, histopathologic characteristics, and treatment responses of children with CNO.

Case report: We carried out a retrospective single-center case series study of pediatric and adolescent patients treated for CNO between 2022-2023 at a tertiary center in Abu Dhabi. Electronic medical records (EMR) were reviewed in order to collect data.

Six patients were included in the study, out of which 83.3% were females. The median current age and age of disease onset was 10.5 years range: 5-14 and 5.5 years range: 4-11.All patients had a pattern of recurrent multifocal disease with bone pain and arthralgia. Sixty-six percent of patients presented with leg (hip, knee) pain, 33.3% presented with limping, 16.6% had back pain, 16.6% had shoulder pain.Mean ESR was 44.6mm/hr.Median CRP was 12 mg/L with 60% of patients with CRP > 5 mg/L. 25% of patients had positive RF and all had negative antinuclear antibodies and negative HLA-B27.The most common affected sites were metaphysis, diaphysis of long bones including tibia 100%, fibula 80%, femur 60%, humerus 60%, thoracic spine 60% and lumbar spine 40%. It was represented on Whole-body MRI (wbMRI) as bone marrow edema and multifocal bone marrow signals—hyperintense signal on STIR and hypointense on T1W. Bone marrow biopsy was done for 2 patients and showed cellular marrow with maturing trilineage hematopoiesis. All patients received treatment with NSAIDs and responded well. Of the patients whose disease was not controlled with NSAIDs, 3 patients received bisphosphonates and achieved remission.

Conclusion: The diagnosis of CNO should include clinical history, laboratory and imaging examination, and histopathological examination. Other causes of chronic bone pain should be ruled out. For treatment, NSAIDs are used as first-line drugs followed by steroids,bisphosphonates, and TNF-α inhibitors. Combination therapy with bisphosphonates and TNF-α inhibitors may be an option for refractory CNO. The limitation of this study is its small sample size. Thus, further studies including more patients from other tertiary centers are required to formulate diagnostic and treatment strategies for CNO.

MIS2023-A-1015

A STAT3 Gain-Of-Function Mutation Presenting with Generalized Lymphadenopathy- A case report

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Introduction: Signal transducer and activator of transcription 3 (STAT3) is a transcription factor that plays an essential role in immune cell proliferation and differentiation. Loss of function (LOF) or gain of function (GOF)

mutations in the STAT3 gene have been linked to immune system dysregulation. Patients may exhibit a wide range of clinical symptoms, making diagnosis difficult in most cases.

Case report: We report a 6-year-old child born to non-consanguineous parents. He was found to have recurrent upper respiratory tract infections (URTI) due to adenoids along with eczema at the age of 2 months. The URTIs resolved after his adenotonsillectomy at 2 years old. Generalized lymphadenopathy was also detected that required him to visit an immunologist, haematologist, and rheumatologist without reaching a definite diagnosis. Lymphoma was suspected but biopsy was consistent with lymphoproliferative disease. Eventually, whole exome sequencing revealed a heterozygous pathogenic GOF mutation in STAT3 gene named c.2144C>T, p.Pro715Leu, which is associated with multiple autoimmune diseases, lymphoma and bronchiectasis. He is currently undergoing bone marrow transplantation for curative treatment of this immune dysregulation disease due to regulatory T cell deficiency.

Conclusion: STAT3 GOF mutation is a novel clinical entity that presents with early onset multiple autoimmunity with lymphoproliferation. The majority of primary immunodeficiencies that were described so far have been linked to LOF disorders. However, more GOF mutations leading to immunodeficiency have recently been discovered. The phenotype's heterogeneity makes it challenging to establish a specific clinical definition. The only reliable diagnostic method currently is genetic analysis. Identifying the genetic cause will change the treatment modalities like targeted immunosuppressive therapy or bone marrow transplantation.

MIS2023-A-1012
Systemic Lupus Erythematosus Onset in a Geriatric Age

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Introduction: Systemic lupus erythematosus (SLE) is a chronic autoimmune disease of unknown cause that can affect virtually any organ of the body. Immunologic abnormalities, especially the production of a number of antinuclear antibodies (ANA), are a prominent feature of the disease. Patients can present with variable clinical features ranging from mild joint and skin involvement to life- threatening kidney, hematologic, or central nervous system involvement. The clinical heterogeneity of SLE and the lack of pathognomonic features or tests pose a diagnostic challenge. To complicate matters, patients may present with only a few clinical features of SLE, which can resemble other autoimmune, infectious, or hematologic diseases. More than 90% of cases of SLE occur in women, frequently starting at childbearing age. The onset of SLE is usually after puberty, typically in the 20s and 30s, with 20% of all cases diagnosed during the first 2 decades of life, with the prevalence of SLE is highest in women aged 14 to 64 years. We are presenting a geriatric woman of 75-year-old that was newly diagnosed with SLE with hematological, renal, and cardiac manifestations.

Case report: A 75-year-old, Palestinian female known case of mitral valve disease, atrial fibrillation (AF), and hypothyroidism. She was on trimetazidine 35 mg bid, atenolol 50 mg bid, furosemide 40 mg daily, oral anticoagulant apixaban 5 mg bid, and levothyroxine 75 mg daily. She presented to the emergency with chest pain, and shortness of breath, for 2 days, together with a cough with no history of fever.

In the emergency room, she was hypotensive (blood pressure 89/54), temperature 36.5°C, HR 106 (electrocardiogram show AF controlled rate). Blood investigations show pancytopenia; white blood cells 1.15 x10(3)/mcL (RR 4-11), absolute neutrophils 0.48 x10(3)/mcL(RR 2-7), HB9.2 gm/L (RR 11.5-15.5), platelets 78 x10(3)/mcL(RR 150-450). Erythrocyte sedimentation rate (ESR) 112 mmHg (RR 0-30), c-reactive protein 4.5 mg/L (RR 0-3), Pro BNP 3902 pg/mL (RR 0-125). On physical examination she was pale with no evidence of malar rash or any skin lesions. By auscultation, the patient had diminished air entry on the left side with a pan systolic murmur at the Mitral valve region. Chest x-ray shows moderate left-sided pleural effusion with underlying lung compressive collapse, marked cardiomegaly. Further blood work showed positive ANA, dsDNA, anti-SSA, and anti-SSB, with low complements (C3, C4) and Urinary 24 hours (hr) protiens of 1,256 mg/day (RR10-149). ECHO showed moderately impaired LV, systolic function EF 40%, global hypokinesia, dilated left atrium, grade II diastolic dysfunction, mitral valve is thickened with a nodule seen in anterior Mitral valve leaflet, there was also severe Mitral regurgitation, effective regurgitant orifice area 0.43 cm^2, regurgitant volume 52 ml, vena contracta 0.8 cm. The regurgitant jet is eccentric, swirling posteriorly, maximum velocity of 4.2 m/s. Proximal iso velocity surface area diameter is 0.8 cm, mild to moderate tricuspid regurgitation. Severe pulmonary hypertension, estimated pulmonary pressure 64 mmHg, no intracardiac thrombus noted, moderate effusion with fibrinous strands, size 1.6 cm inferiorly, and pleural effusion noted. Based on the clinical presentation, and laboratory findings, a diagnosis of SLE had been given.

The case was started on pulsed steroids for 3 days one gm daily for 2 successive days, followed with oral prednisolone one gm/kg then tapered gradually, together with hydroxychloroquine 200 mg bid and mycophenolate mofetil 500 mg bid that was escalated gradually to 2 gm daily. She was shifted from oral apixaban to warfarin with a target International normalized ratio of 2-3.

The case was reviewed after that in follow-up visits, show marked clinical improvement, a repeat of ECHO showed marked improvement in the mitral valve regurgitation from severe degree to mild to moderate mitral regurgitation. Repeated ECHO showed also improvement in LV function.

Conclusion: Systemic lupus erythematosus can present for the first time at an elderly age. The first presentation can be a cardiological manifestation with Libman-Sacks endocarditis, pancarditis affecting the tricuspid valve and LV function, pericarditis, and pleural effusion.

MIS2023-A-1025

ANCA Positive Vasculitis- Rheumatoid Arthritis Overlap with Enthesitis Responding To Rituximab Followed by Upadacitinib Therapy

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Introduction: A 41-year-old Emirati-Arab female was admitted with 8 weeks history of severe cough and dyspnoea which failed to respond to oral and IV broad spectrum antibiotics after initially diagnosed as chest infection. She developed red painful eyes, fatigue, painful wrists and ankles, and loss of sensation in right ulnar nerve distribution. Her c-reactive protein (CRP) was very high (647 mg/L), anemia (hemoglobin [Hb] 8.1 g/dL, mean corpuscular volume 58 fL), thrombocytosis (648 cellsx109 /L), She has a family history of psoriasis (brother), rheumatoid arthritis (sister), and celiac disease (brother). Eyes examination revealed features suggestive of vasculitis scleritis. High-resolution computed tomography showed multiple bilateral patchy alveolo-interstitial infiltrates in both

lungs involving both upper lobes, lower lobes and right middle lobes, some showing tree in bud appearances associated with ground glass attenuation and associated septal thickening. Nerve conduction confirmed features of right ulnar nerve mononeuritis multiplex.

Case report: Sonographically, she had moderate synovitis in right second metacarpophalangeal joint with erosion, tenosynovitis of 1st and 6th extensor compartments in the wrist and right Achilles enthesitis. Blood test revealed positive c-ANCA PR3 (1:1000), rheumatoid factor (RF) (92 mg/L), negative CCP, positive antinuclear antibody (ANA) (granular)(1:320), positive anti-double stranded DNA (dsDNA) antibodies (1:100). Normal C3 (1.76 g/L) and mildly raised C4 (0.49 g/L), normal ferritin (66.0 ug/L), mild raised urine protein (206 mg/ L (60-140)) with normal creatinine and liver function. Patient has no tissue amenable to biopsy. Based on clinical and serological finding patient was diagnosed as ANCA positive vasculitis with evidence of pulmonary, ophthalmic and nerve vasculitis and arthritis. Patient had erosive polyarthritis with positive RF suggestive of rheumatoid arthritis and significant enthesitis with strong family history of psoriasis. Patient was ds-DNA positive but no features of lupus.

Patient was given prednisolone 40mg daily with gradual tapering and had 2 infusions of Rituximab 1000mg IV 2 weeks apart. After which she has improved significantly within 4 weeks with complete resolution of chest symptoms, eyes signs and symptoms and dropping of CRP <4 and urine protein to <60 mg/L and Hb improved to 11.7 g/dl and platelets to 445 cellsx109 /L with resolution of chest radiography findings. She continued to stay to be c-ANCA PR3 and dsDNA positive after rituximab therapy. Ulnar mononeuritis multiplex symptoms completely resolved after 4 months. Patient was initiated on azathioprine 100 mg (2 mg/kg) which caused mild neutropenia and subsequently dose was dropped to 50 mg daily. Patient however continued to suffer from right Achilles tendon enthesitis pain and ultrasound showed left Achilles tendon severe enthesitis with erosions, calcification, enthesophyte, abnormal tendon echogenicity and thickness and severe power Doppler signal and retrocalcaneal bursitis. In the right side there was moderate enthesophytes and features of enthesitis and mild bilateral plantar fasciitis. Patient started to have 13 tender joints and 4 swollen joints 8 months after last dose of Rituximab with no flare of pulmonary, neurological, renal, ophthalmic vasculitis features. Patient was switched to upadacitinib and improved significantly and achieved 2 swollen joints within 3 months and no swollen or tender joints after 6 months with resolution of all enthesitis. She continued maintenance of upadacitinib 15 mg and azathioprine 50 mg combination. c-ANCA became negative after Upadacitinib treatment. Patient reported weight gain of 5 kg since starting Updacitinib.

Conclusion: Rare cases of overlap of ANCA positive vasculitis and rheumatoid arthritis has been previously reported, however the combination of that with polyenthesitis and dsDNA-positivity was unique. The response of the vasculitis and arthritis components but no the entheistis to Rituximab was probably due to different autoimmune drive independent off B cells, given the strong psoriasis family history and might indicate poliautoimmunity with autoimmunity activity at different pathways simultaneously. The effectiveness of JAK inhibitor (upadacitinib) as a maintenance therapy in this case was of special interest and might guide treatment in future patients with similar overlap syndrome.

MIS2023-A-1032
ISG15 Deficiency: the Diversity of Interferon Responses

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HIGHLIGHTS FROM MIDDLE EAST IMMUNOLOGY SUMMIT OF THE GCC AIR

Introduction: TISG15 deficiency is an ultra-rare disease that was recognized over a decade ago. Since then, only a few cases have been reported worldwide. It was described as a mixed syndrome of Mendelian susceptibility to mycobacterial disease and monogenic interferonopathy.

Case report: We report a boy with ISG15 deficiency presenting with migratory arthritis. The patient's background was suggestive of immune dysregulation including bronchial asthma, atopic dermatitis, mild recurrent skin lesions, recurrent admissions with pneumonia and hypoxia, and failure to thrive. Further, his family history was significant for consanguineous parents, a brother with a history of BCGitis and hypogammaglobulinemia, and a cousin with recurrent infections, severe skin lesions, and multiple food allergies. The patient was investigated for immune dysregulation, whole exome sequencing revealed homozygous mutations in ISG15 and COL9A3 genes. Family segregation showed homozygous mutation in the brother who had BCGitis and heterozygous mutation in the parents and sister who are asymptomatic. Further, brain MRI of the patient confirmed subcortical and globus pallidus calcification, which is described in individuals with ISG15 deficiency, secondary to enhanced INF-I. Positive serology of ANA and anti-smith antibodies were detected. The patient was relatively well, apart from recurrent fevers and migratory arthritis flares.

Conclusion: This report introduces a novel presentation of ISG15 deficiency with a combination of autoinflammation and autoimmunity, and highlights the contrasting functions of this gene contributing to its variable phenotypic expressivity.

MIS2023-A-1050

Sensorineural Hearing Loss Precede the Onset of Psoriasis

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Introduction: Psoriasis (PsO) is a common chronic inflammatory skin disease that may exhibit a variety of clinical manifestations. Psoriasis has also been identified as a multisystem chronic inflammatory disorder associated with multiple comorbidities. A few studies have reported that PsO can also be associated with sensorineural hearing loss (SNHL) since both diseases may share an immune-mediated and systemic inflammatory pathogenesis, yet the clinical significance remains undefined, and the extent of association has not been well defined. Immunologically, it has been explained as an interaction between circulating antibodies and an antigen in the inner ear, or by direct inner ear by activated T cells.

Case report: A 62-year-old Iranian male patient. At the age of 20, was diagnosed as a case of Left profound SNHL, right moderately severe to profound SNHL. Since then, hearing aids have been used by patients. At the age of 55, he had been referred from the ear-nose-throat clinic to the Rheumatology Clinic of Al Kuwait-Dubai (Al-Baraha) hospital with red, itchy, scaly skin rash in the groins, genitalia, and the chest. Moreover, there was pain and swelling of the whole left thumb with redness.

Physical examination showed psoriatic patches over the extensor surface of the forearms, lateral aspects of the thighs, shins and back of the hands. There was swelling of the whole left thumb; sausage shaped (dactylitis), with redness and tenderness. Blood investigations revealed normal complete blood count, liver function test, renal

function test, erythrocyte sedimentation rate; ESR, and c-reactive protein. Rheumatoid factor was negative. In view of skin psoriasis and thumb dactylitis, He was diagnosed with psoriatic arthritis and PsO.
Methotrexate had been started, and in 2 months review there was remnant redness with little of silvery scales over the right groin and around the umbilicus, with improvement of the dactylitis.

Conclusion: In PsO, SNHL might precede musculoskeletal manifestations.

MIS2023-A-1049
Toxic Epidermal Necrolysis in Concurrence with Psoriasis

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Introduction: Toxic epidermal necrolysis (TEN) is a severe blistering disease. Although the exact pathophysiology is not well known, the possible role of Th17 cells in the pathogenesis of TEN was reported. Th17 cells help with the recruitment of neutrophils and other inflammatory leukocytes with inflammation and tissue damage. On the other hand, Th17 cells are also intensely involved in the pathogenesis of psoriasis (PsO); chronic papulosquamous skin disease, and pustular psoriasis.

Case report: A 40-year-old woman, of Indian ethnicity, with psoriasis for 10 years, on local treatment, presented to the accident emergency department of A Kuwait-Dubai hospital (Al Baraha), Dubai, United Arab Emirates with a 5-day history of fever, followed by a generalized eruption of skin boils that rupture quickly with peeling of the skin, and discharge thick exudate materials. Interestingly she had a similar episode 2 years before but with less severity. On physical examination, vital signs were stable, generalized skin redness of the body, erosions, exfoliative, peeled skin with scaly crusts spread, widely over the body trunk and extremities, involving more than 70% of the body surface area, with Foully smelly skin discharge, and positive Nikolsky sign. Oral mucosa, conjunctiva, and genitalia areas were normal.

Laboratory investigations revealed high white cell counts (WCC) 29.7 (RR: 4.80-10.10x103/ul, with a normal differential WCC count, high inflammatory markers; erythrocytes sedimentation rate (ESR) 111 mm/hr (RR 10-20), and c-reactive protein 50 mg/dl (normal <0.3 mg/dl), and low Albumin 25.5 g/l (RR: 34-50). Electrolytes and renal function showed impairment in the renal function with low sodium (NA) Na 131 mmol/l (RR: 13-145), high potassium K 5.2 mmol/l (RR: 3.4-4.7), uric acid 538 umol/l (RR: 155-357), creatinine 139 umol/l (RR: 49-90), and urea 18.3 mmol/l (RR: 2.50-6.40). Her SCORTEN score was 4 points (age ≥40 years, Detached or compromised body surface >10%, serum BUN >28 mg/dl (10 mmole/l), and blistering body surface ≥10%. Skin swabs demonstrated staphylococcus intercedes. Skin biopsy revealed increased dermal vascularity with per vascular chronic mononuclear inflammatory cellular reaction that focally invade blood vessel walls. Epidermal mild hyperkeratosis and parakeratosis are observed. Rete ridges are slightly elongated in focal areas while it was normal in others. Few polymorphs are detected at the hyperkeratotic surface area and focally at the dermis.

Supportive care was applied including admitting the patient to the isolation room, fluid and electrolyte balance, nutritional support, pain management, and protective dressings with topical miconazole, emollient, and sodium fusidate ointment, and cetirizine 10 mg OD. She stayed in eh hospital for 9 days. At the time of discharge, the body

was clear, fully regenerating skin epithelium, with no more skin eruption or exfoliation. And, blood investigations were all back to normal

Conclusion: Toxic epidermal necrolysis can occur in concordance with PsO.

MIS2023-A-1048
Tocilizumab as an Effective Treatment for Non-Infectious Scleritis in Rheumatoid Arthritis

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Introduction: Scleritis is a serious painful inflammatory eye disease that might result in visual loss. Around 50% of scleritis patients have an underlying systemic autoimmune disease of which rheumatoid arthritis (RA) contributes to the majority. Tocilizumab is an Interleukin 6 (IL-6) inhibitor biological medication approved to treat adults with moderately to severely active RA, with some case series that it might be effective in treating eye scleritis in RA patients. But not no case report demonstrated a positive response of scleritis to tocilizumab in RA patients with failure to multiple Biological disease-modifying anti-rheumatic drugs (bDMARD).

Case report: A 67-year-old Indian woman who has suffered from RA for 5 years initially treated with Methotrexate 25 mg Qwk and Sulphasalazine 1 g TID, with no response. Followed by Adalimumab (anti-tumor Necrosis factor; anti-TNF) 40 mg a week. Rheumatoid arthritis musculoskeletal symptoms and signs, and the laboratory inflammatory markers responded to adalimumab. But, despite the response to adalimumab, she developed left eye non-infectious anterior scleritis that had been confirmed by an ophthalmologist. The scleritis was active and persistent, despite adding systemic corticosteroid. Therefore, Adalimumab was changed to Etanercept (anti-TNF), which has been used for 6 months. Again, there was a good MSK response and normalization of the inflammatory marker to Etanercept with no improvement in the scleritis. After which, Etanercept had been changed to Rituximab; a monoclonal antibody (anti-B-lymphocyte antigen CD20). Scleritis responded to the Rituximab with complete resolution of the scleritis and improvement in visual acuity. The response to Rituximab continued for 3 years. After which, the scleritis recurs again. The patient started to complain of a reduction in the visual acuity. In an attempt to hinder the scleritis and the inflammation in the eye. Tocilizumab had been started. One month after starting the Tocilizumab the redness of the eyes improved and the patient started to have visual acuity improvement.

Conclusion: Tocilizumab might be an alternative treatment for RA patients with scleritis failing other biological treatments. Long-term studies are required to establish the safety and efficacy of Tocilizumab in treating non-infectious scleritis related to RA.

MIS2023-A-1047
Lipodystrophy and Calcinosis; Two Rare Manifestations in Adult Dermatomyositis

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HIGHLIGHTS FROM MIDDLE EAST IMMUNOLOGY SUMMIT OF THE GCC AIR

Introduction: Dermatomyositis (DM) is an autoimmune connective tissue disease (CTD) characterized by special skin rashes and muscle inflammation. Calcinosis occurs as a complication of DM due to abnormal deposition of insoluble calcium salts in tissues, including skin, subcutaneous tissue, fascia, muscle, and tendons. Although calcinosis is more common in juvenile DM (JDM), it can occur in adult DM. Muscle calcification is generally asymptomatic, and may be seen only on radiological assessment. Calcinosis often correlates with the level of long-term disability. It is usually a sign of "burned-out" myositis; it contributes to muscle atrophy and joint contractures. On the other hand, lipodystrophy (loss of subcutaneous fat) is another rare manifestation characterized by loss of adipose tissue that can be seen in 10% of primarily JDM, and very rarely in adult-onset DM.

Case report: A 39-year-old Sudanese case of DM for 10 years and mild bronchial asthma for 8 years (not on regular treatment). Mrs. M.A.T presented to Al Kuwait-Dubai hospital (Al-Baraha) for the first time with fever, lower back pain, body rash, increase in hair loss, mouth dryness, and blurring of vision. But there was no muscle weakness. The symptoms were of a week duration. The medications she was on were esomeprazole 40 mg OD, Prednisolone 5 mg OD, Azathioprine 200 mg BID, diltiazem hydrochloride 180 mg OD. On examination: vital signs were normal. Eyes were surrounded with heliotrope rash, mechanic's hands. There were regional localized areas in the back and over the frontal aspect of the right thigh with loss of subcutaneous fat, and dark discoloration of the skin.

Laboratory work revealed: Negative rheumatoid factor (RF), anti-dsDNA, anti SS-A/RO, anti SS-B/La, anti-RNP, anti-centromere antibody, anti-JO1, anti smith antibody (AntiSM), anti-scleroderma 70, anti-histone, anti-cardiolipin antibody. Complete blood count showed normal total and differential white cells and platelet counts, but slightly low hemoglobin at 11.1 g/dl (RR: 10.5-13.5), with mean cell volume of 75.9 fl (RR: 71.0-90.0), and mean cell hemoglobin of 25.5 pg (RR: 24.0-29.0). Associated with low serum iron at 46 ug/dl (RR: 60-160), low serum ferritin at 11.6 ng/ml (RR: 13.0-150.0), and normal serum transferrin at 264 mg/dl (RR: 192-382). There were no significant changes in the level of the muscles enzymes: Myoglobin <21 ng/ml (RR: 25-72), creatine phosphokinase (CPK) 60 U/l (RR: 39-400), Creatine Kinase MB (CK-MB) 6.3 U/l (RR: 2.3-9.5), Troponin T hs STAT 0.004 ng/ml (RR: 0.013-0.025), and lactate dehydrogenase 509 U/l (RR: 280-500) Inflammatory markers showed a high erythrocyte sedimentation rate (ESR) of 34 mm/hr. (RR: 1-20), and normal C-reactive protein (CRP). Radiological examination showed a pelvis x-ray with small focal globs of amorphous calcification in the muscles. Lumbar and knee x-rays with osteoarthritis changes. Methotrexate 20 mg/week was started. She was prescribed alendronate 70 mg/wk, CaHco3 500 mg TDS, vitamin D (1 Alfa) 0.25 microgram OD (as DEXA showed osteopenia). Iron fumarate 200 mg BID, and artificial tears and saliva. In 6 months, prednisolone and omeprazole stopped, Azathioprine was reduced to 50 mg OD. This case responded well to medications, with no clinical symptoms, and normal muscle enzymes and inflammatory markers.

Conclusion: Both calcinosis and lipodystrophy can be seen in the adult dermatomyositis.

MIS2023-A-1046
Behcet's Disease and Psoriasis Arise Together

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Introduction: Behcet's disease (BD) is an inflammatory systemic vasculitis condition of unknown etiology, involving

multiple organ systems such as mucous membranes, skin, joints, intestines, lungs, central nervous system, and vessels. Psoriasis is a chronic proliferative and inflammatory condition of the skin, characterized by erythematous plaques covered with silvery scales. Despite that the pathogenesis of BD and psoriasis share common perspectives, yet, reports of patients who have both diseases are rare. Therefore, BD may place individuals at increased risk of PsO. The risk had been suggested to be more in the male cohort, and senile population. On the other hand, the inclusion of Behcet's disease among seronegative spondyloarthropathies is still being debated.

Case report: A 48-year-old Arab man was referred to rheumatology from the ophthalmology clinic (where he is under care for recurrent bilateral iritis). Detailed history revealed a history of chronic sinusitis, Recurrent rash over the shins and the extensor surface of the forearms, recurrent painful mouth ulceration, recurrent genital ulceration, Headache on and off for the last few years, chronic constipation, generalized bone pain, and low back pain. The lower back was typical of inflammatory back pain in characteristics. Family history was significant for diabetes in the father and mother, hypertension and cardiovascular disease in the father, asthma in the mother, and systemic lupus erythematous in a paternal cousin. Examination revealed a small macular rash over the anterior surface of the right shin, and the extensor surface of the left elbow surface of the right shin. Also, interestingly there were dry, itchy, raised skin patches (plaques) covered with white silvery scales over the extensor surface of the right knee. Laboratory investigations revealed normal complete blood counts, erythrocyte sedimentation rate (ESR), C-reactive protein, liver, and renal function. Antibodies profile and HLA-B27 were all negative. X-rays of the cervical, thoracolumbar, and sacroiliac joints (SIJs) came normal. Magnetic resonance imaging for SIJS was completely normal with no evidence of edema or erosions. The diagnosis was BD based on the International Criteria for Behcet's Disease (ICBD), and skin PsO based on the presence of a psoriatic lesion in the clinical examination. Whether the BD and the skin PsO are part of a broader spondylarthritis (especially in view of the history of inflammatory back pain) is not clear as the symptoms do not fit within the Spondylarthritis International Society (ASAS) criteria.

Conclusion: Behcet's disease may be associated with a significantly increased risk of psoriasis, and probably psoriatic arthritis.

MIS2023-A-1041
Adalimumab Cause Hidradenitis Suppurativa in Psoriasis Patient

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Introduction: Hidradenitis suppurativa (HS) is a chronic, inflammatory skin disorder that affects apocrine units. Psoriasis is a chronic inflammatory disorder that prominently affects the skin and joints. Although the relationship between psoriasis and HS has been reported in a number of case reports, yet, some evidence is controversial. A systemic review reported while a minority of patients had HS before psoriasis, others suffered from paradoxical HS following biological therapy used to treold Syrian patient with psoriasis for 7 years, on infliximab treatment. Seen for the first time by the rheumatology service when she was admitted to the medical ward of Al Kuwait-Dubai Hospital (Al Baraha) for severe flare-up of skin psoriasis (PsO) accompanied by the appearance of psoriatic arthritis (PsA) the first time, after stopping taking the PsO treatment (Infliximab) for 7 months, when she was in process of moving from Syria to the United Arab Emirates (UAE).

Physical examination proved the presence of multiple joint arthritis (involving elbows, proximal interphalangeal joints, knees, and ankles). The body was full of skin psoriasis. Infliximab had been resumed and the condition came under control with remission of PsO and PsA. A few months after, MM was shifted to Adalimumab as part of the patient support program. Three monthly assessments revealed a flare-up of PsO, and nail PsO. As well, there were areas of scars with depression at different places on the body. MM stated that since she started on Adalimumab, she had recurrent boils that required surgical excision. The boil excision was followed by an area of depression and scars at the place of excised boil. The diagnosis of the recurrent boils was an HS.

As a result of failure to Adalimumab, MM was shifted back to Infliximab. One week after the Adalimumab there was a complete clearance of the skin PsO. After 5 years of follow-up, PsO, and PsA were all in remission, with no recurrence of boils.

Conclusion: The cytokine profiles of HS, psoriasis, and anti-TNF-induced psoriasiform lesions are very similar. Therefore, like psoriasis, HS is a rare skin lesion that can develop while on anti-TNF therapy and is perhaps due to cytokine dysregulation as has been seen with TNF antagonists. The cytokine dysregulation that results in HS is class-specific (Adalimumab).

MIS2023-A-1044
Rheumatoid Nodules may Precede Onset of Clinical Rheumatoid Arthritis

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Introduction: Rheumatoid nodules are the most common cutaneous manifestation of rheumatoid arthritis (RA) that can be seen in about 25% of RA patients and are found mostly in subcutaneous regions subject to recurrent mechanical stress. On the other side, the ganglion cyst starts when the fluid leaks out of a joint or tendon tunnel and forms a swelling beneath the skin. The cause of the leak is generally unknown but may be due to trauma or underlying arthritis. The key difference between an RA nodule and a ganglion cyst is that a ganglion cyst is a small, soft, fluid- filled lump that occurs under the skin and is not associated with any medical condition, while an RA nodule is a large, firm lump that occurs under the skin and is usually associated with RA. Rheumatoid nodule is most often seen in patients with seropositive RA and more severe disease. However, there is no evidence that systemic therapy treats rheumatoid nodules, and treatment of rheumatoid nodules is often not necessary.

Case report: A 60-year-old Indian male, who is not known to have any medical problems before. Presented to the orthopedic clinic with a small mass at the dorsum of the right hand that appeared over a few days. The mass was a lump of 3x2 cm with a defined border, mobile, and no tenderness. The skin over the cyst looks normal and feels smooth, round, and rubbery. Despite the small mass of the cyst and the presence of no symptoms, the patient wanted to remove the mass for cosmetic reasons.

Excision was done surgically and the mass was sent for histological examination as a routine practice in the hospital for any surgically excised sample. The histology results revealed macroscopically a specimen consists of multiple grey-white soft tissue bits altogether measuring 3x2x0.5 cms

Microscopic examination showed a section with synovial tissue with villous hyperplasia of synovium. The villi are lined by multilayered synoviocytes. There were areas of fibrinoid necrosis surrounded by a palisade layer of

histiocytes. The stroma shows extensive vascular proliferation with a focal collection of lymphocytes. Occasional fragment shows fibrinoid necrosis of the vessel wall with lymphoid aggregation, as well as areas of fibrosis. The histology report concluded with the impression that histopathological features are suggestive of RA nodules.

Based on the histological findings the patient had been referred to the rheumatology clinic of Al Kuwait-Dubai (Al Baraha) hospital for further evaluation for any possible clinical RA. Detailed history and physical examination did not reveal any musculoskeletal manifestation. Laboratory tests revealed normal complete blood count, inflammatory markers (erythrocyte sedimentation rate and c-reactive protein), and, renal and liver function. The immunology profile revealed negative rheumatoid factor and anti-citrullinated peptide, antinuclear antibody, (ANA) and extractable nuclear antigen (ENA). Based on the absence of evidence to treat rheumatoid nodules the patient was discharged with advice to attend if any new symptoms emerge.

After 13 months, the patient presented again with typical swelling and pain in the joints of the fingers. Examination revealed arthritis (swelling, hotness, and redness) of metacarpophalangeal joints (MCPJs) and proximal interphalangeal joints. Squeeze tests were positive with a tenderness of MCPJs. Repeated blood tests showed normal complete blood count, and renal and liver function. The immunology profile revealed negative rheumatoid factor and anti-citrullinated peptide, ANA, and ENA. But, the high ESR is 65 mm/hour, and the CRP is 42 mg/dl. The patient was diagnosed with RA based on the American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) criteria for RA classification 2010. Although Methotrexate could increase the rheumatoid nodule, it was the first option of treatment. 15 mg Qwk had been started. Assessment in 3 months the patient came back with no joint pain, swelling, or tenderness, with normal inflammatory markers.

Conclusion: A case of an asymptomatic patient with a dorsum subcutaneous nodule that had been suspected of as ganglion. Histology proved the mass an RA nodule. Rheumatoid arthritis symptoms followed after years.

MIS2023-A-1042
Temporary Raynaud's Phenomenon Secondary to COVID Vaccination

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Introduction: Raynaud's phenomenon (RP) is a condition that causes the blood vessels in the extremities to narrow with restriction of the blood flow. RP usually affects the fingers and toes but rarely might occur in the ears or nose. Secondary RP occur usually due to the presence of underlying rheumatological autoimmune disorder, and to a lesser extent due to coagulation disorders. While Coronavirus disease-19 (COVID-19) originated in Wuhan-China, within a few months it was declared a global pandemic by the World Health Organization (WHO), and a number of vaccines received emergency approval in an attempt to compact the COVID-19 that is highly contagious. RP in association with COVID-19 vaccination is generally rare. Few cases were reported. No previous report came from the Middle East region.

Case report: A 32-year-old, Arabic woman presented to the virtual clinic with transient attacks of well-demarcated, whitening fingers, of both hands which were triggered by exposure to cold environment and accompanied by a sensation of numbness. The patient sent a photo to explain her complaints. The photo revealed well- demarcated white fingers on the right 2nd, 3rd, and 4th finger, and on the left 3rd, 4th, and 5th finger. She was given an urgent

appointment the next day at the clinic. On presentation to the clinic, she gave a history of receiving her second injection of the inactivated SARS-CoV-2 vaccine, BBIBP-CorV one week prior to the development of the RP. Ms. KA is otherwise healthy with no other comorbidities or autoimmune diseases, she has no known allergy, is not married, and not using oral contraception or any other medications. She is not a current or ex-smoker. And could not recall any vibratory triggers. She works as an HR personnel with no previous history and no family history of RP.

On examination, there were well-demarcated, white-pale, cold areas involving the volar and dorsal aspects of the right 2nd, and 3rd finger, and the left 2nd, 4th, and 5th finger. The remaining digits showed no abnormalities There was no digital pitting, ulceration, or gangrene. The peripheral pulses of both arms were symmetrical and of normal character.

Investigations revealed: normal complete blood count and biochemistry, coagulation parameters including cryoglobulins. Antibodies profile was all negative for ANA, Anti-dsDNA, anti SS-A/RO, Anti SS- B/La, Anti-JO1, Anti smith antibody (AntiSM), Anti-RNP, Anti-centromere antibody, anti-scleroderma 70, anti-Histone, Anti-cardiolipin antibody, rheumatoid factor (RF), and anti-CCP. Normal complements and cryoglobulins range. Antibodies to the heparin/platelet factor 4 complex are not detectable. coagulation profile was normal including antibodies to the heparin/platelet factor 4 (PF4) complex.

The absence of current or previous immunological or coagulation disease, the absence of RP in her past medical history, the lack of any RP risk factors and triggers, complete normal related blood tests, and the recent administration of the COVID-19 vaccine collectively raise doubt about RP relation to the inactivated CVOID-19 vaccine.

Conclusion: We reported a case of a new-onset RP after COVID 19 vaccine, which occurred in an otherwise healthy young Arabic ethnicity woman, who lacked any known risk factors and associations with possible causes for secondary RP.

MIS2023-A-104

A Challenging Case: When Lupus and ANCA Antibodies are Positive with AKI and a Catastrophic Biopsy

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Introduction: It is difficult to diagnose lupus nephritis and ANCA vasculitis based on positive antibodies alone, kidney biopsy is very important. The overlap of rheumatoid arthritis and systemic lupus erythematosus is well described, with a syndrome known as 'rhupus'. ANCA-associated vasculitis, however, is uncommonly associated with other autoimmune conditions. It is rare to have overlap between lupus nephritis and ANCA vasculitis, many case reports mentioned about C-ANCA and lupus nephritis coexisting but nothing was mentioned about C-ANCA and P-ANCA with positive lupus markers.

Case report: A 69-year-old female, is known to have diabetes mellitus, hypertension, and dyslipidemia. Of note, she had a history of covid 19 infection in 2019 and hip joint replacement in 2022. She was diagnosed as having Systemic lupus erythematosus and started on Mycophenolate mofetil 500 mg twice a day and oral Prednisolone with +ve cANCA and pANCA she underwent kidney biopsy complicated with massive bleeding required embolization,

unfortunately, the biopsy was inadequate. the patient's renal function continued to deteriorate reaching end-stage renal disease and she was started on hemodialysis. Other medical problems included pancytopenia with high kappa/lamba, and polyclonal gammopathy, suggested to be related to viral or hydralazine despite that, Anti Histone AB was negative.

The patient stabilized her creatinine to the range of 250-350 micromol/L however she is still dialysis dependent despite having urine production >500 ml/day. her immunosuppression medications initially included intravenous pulse steroids and mycophenolate mofetil dose of 1500 mg twice daily, and it was reduced gradually to 500 mg bid after her spinal abscess.

Conclusion: Complex cases require complex investigations, but when kidney biopsy is inadequate and complicated, treatment is very difficult and the balance between immunosuppression and prevention of opportunistic infections is very difficult.

MIS2023-A-1037

Hyper-Eosinophilic Syndrome in Hemodialysis Patient Revealed a Challenging Diagnosis

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Introduction: Eosinophilic granulmatosis with polyangiitis (EGPA) is a small vessel vasculitis characterize by asthma, peripheral eosinophilia, and various degrees of skin, renal and neurological manifestations. The serology for P-ANCA or anti-myeloperoxidase is positive in 50% of cases. We describe very rare association of PR3-ANCA in EGPA diagnosed in hemodialysis patient.

Case report: A 63-year-old female, known to have long-standing hypertension >25 years. She had ESRD and started on hemodialysis (HD) via right IJ PermCath on 5/2019. She developed chronic hypereosinophilia with symptoms of dyspnea, rash, fatigue and nausea during HD. She also had abnormal MIBI scan EF 39%, ischemic changes suggested but no cardiology follow up. Overtime, she also developed leukocytosis mainly eosinophilia and thrombocytopenia. She was diagnosed with HD membrane allergy (allergy type 1) on 10/2021 (F7, & PMMA membrane). The eosinophilia improved and thrombocytopenia resolved after changing HD membrane and using steroid. She was admitted several times for HD PermCath malfunction and fever with unclear focus, or pneumonia/reactive airway disease. On 4/2022, she was admitted for evaluation of 1-week symptoms of fever, wheezes, left sided chest pain/severe back pain, dizziness and arthralgia. The investigations revealed eosinophilia 2.75 x10^9/L, IgE: 391.0 IU/mL, ESR: 54 mm/hr, and PR3 C-ANCA 129 RU/mL. CT chest and Echo showed large pericardial effusion and she underwent pericardial drain (1.5 L of bloody fluid with negative cytology & TB). She was diagnosed with PR3-ANCA associated EGPA and treated with steroid and azathioprine. She had favorable outcomes with improvement in cardiomyopathy and no allergic symptoms.

In a retrospective European multicentre cohort study including 845 EGPA patients, (28.6%) patients had MPO-ANCA and only 16 (2.2%) were PR3-ANCA positive. PR3-ANCA EGPA patients share clinical features with granulomatosis with polyangiitis (less asthma, peripheral neuropathy and more cutaneous manifestations, pulmonary nodules, lower median eosinophil count). In review of literature, only one case reported cardiac

tamponade with PR3- ANCA EGPA vasculitis. Our case had PR3 ANCA EGPA vasculitis in HD patient with HD membrane allergy, and cardiomyopathy with hemorrhagic pericardial effusion.
Conclusion: Onset of EGPA vasculitis in hemodialysis patient is uncommon and the associated with PR3-ANCA and hemorrhagic pericardial effusion is very rare. Prompt and early diagnosis vasculitis and initiation of steroid and other immunosuppression medications yielded favorable outcomes. Evaluation of hyper-eosinophilia in dialysis patients should include dialysis membrane allergy, workup of vasculitis and exclusion of other causes.

MIS2023-A-1033

The coexistence of Systemic Sclerosis, Systemic Lupus Erythematosus and Celiac Disease. An Unusual Combination of Multiple Autoimmune Syndrome. A Case Report

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Introduction: Multiple autoimmune syndromes (MAS) involves the coexistence of multiple autoimmune disorders due to immune dysregulation pathways and shared genetic polymorphisms. Systemic sclerosis (SSc) and systemic lupus erythematosus (SLE) are connective tissue diseases, with different and unique pathogenesis and antibody profiles. Celiac disease (CD) is an immune-mediated small intestine pathology. the coexistence of SLE and CD, SSc and CD, and SSc and SLE (also known as overlap syndrome) has been reported previously in some case reports.

Case report: A 21-year-old woman with juvenile SSc and interstitial lung disease (ILD) was admitted to the hospital in January 2023 with a history of fever and cough with an impression of upper respiratory tract infection. She reported persistent lower abdominal pain, diarrhea, and weight loss over the past three months. Laboratory investigations revealed leukopenia, normocytic normochromic anemia, and thrombocytopenia, after investigating the causes of thrombocytopenia, labs reported positive ANA (antinuclear antibody) and anti-DsDNA (anti double Stranded DNA) . The patient was diagnosed with Systemic Lupus erythematosus and started on steroid and hydroxychloroquine. An upper GI endoscopy done and biopsy was taken which revealed intraepithelial lymphocytic infiltration and villous atrophy that led to requesting Celiac serology, which came back positive for Antideaminated gliadin peptide (DGP) IgA and tissue transglutaminase (TTG) IgA, the diagnosis of Celiac disease (CD) was, hence, made. The patient was educated about the coexistence of SLE and CD in addition to SSc and advised to follow a gluten-free diet, hydroxychloroquine, mycophenolate mofetil, and prednisolone. A follow up visit was conducted six months later, the patient's condition stabilized, with resolution of her fever and pancytopenia, abdominal pain and diarrhea resolved as well, and patient continued to gain weight.

Our patient has a unique presentation of CD and SLE that develops 9 years after the onset of SSc. Patients with a history of one autoimmune disease Have roughly a 25% Chance to acquire an additional type of autoimmune pathogenic disorder. Few case reports discussed the coexistence of CD in SSc patients as well as the association between CD and SLE. The association between SLE and SSc is well- established in literature. Despite these past correlations in case studies, no previous case reports documented the coexistence of SSc, SLE, and CD together.

Conclusion: To the best of the author's knowledge, this case study is the first to report multiple autoimmune syndromes coexisting in a single patient, including SLE, SSc, and CD. The combination fits the criteria of MAS but cannot be classified into any subcategory of the already known MAS types.

This case was reported to highlight the importance of investigating the coexistence of autoimmune syndromes based on clinical presentation, as rare associations can occur.

MIS2023-A-1030

More Than Meets the Eyes – A Case Report on Rare Cause of Extensor Tenosynovitis

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Introduction: Tenosynovitis, inflammation of fluid-filled synovium within tendon sheath, could be secondary to arthritis (rheumatoid arthritis, gout, spondyloarthritis, such as), overuse, infective (post injury, bite or spread from other sites) and idiopathic. Reactive tenosynovitis secondary to distant infection is a rare phenomenon. Untreated chronic tenosynovitis can result in finger stiffness, deformities, adhesions and contractures. To rule out any treatable pathology is of astute importance in such cases. Here we describe case of a common infection with uncommon presentation as bilateral reactive extensor tenosynovitis of forearm.

Case report: A 54 year male, visited Rheumatology Out Patient Department with persistent swelling of bilateral forearms for 3 years. He denied history of any joints pain/joint swelling, skin rash, low back pain, fever, cough, weight loss, diarrhea or bleeding per rectum, any other significant rheumatic or other systemic illnesses. On examination, he had lobular, fluctuating, non-tender, non-pulsatile swelling over bilateral forearm extensor aspect. On review of records, he had persistently high inflammatory markers with normal hemogram, liver and renal function. His rheumatoid factor, anti CCP, ANA and HLA B 27 were negative. His previous ultrasound scan of bilateral forearm showed fluid collection with diffuse thickening of sheath of extensor digitorum and extensor carpi radialis longus and brevis tendons in forearm extending upto wrist. Bilateral wrist and other small joints were normal. He had previously been treated with multiple courses of NSAIDs and oral steroids, with minimal to no response.

Ultrasound guided aspiration of collection around extensor digitorum tendon was done and analysis revealed inflammatory fluid with total leucocyte count 17,450 (with 70% polymorphs). Gram's stain, AFB stain, Tb-PCR, crystal analysis and culture of same were negative. On further work up, his Mantoux was 18 mm and Tb-quantiferon was positive. He, however, had no current symptoms/past history or contact with active tuberculosis patient. His chest x-ray was unremarkable, however HRCT chest showed multiple tiny well defined centrilobular nodules in bilateral lungs with few enlarged pre-vascular lymph nodes. His sputum AFB, Tb-PCR, gene-expert and culture were negative. He denied any invasive intervention. Hence, under high clinical suspicion of tuberculosis with reactive bilateral extensor tenosynovitis, he was started on anti-tubercular regime. He received intra-tendinous steroid injection over all peritendinous collection. On follow up, his CRP showed significant reduction with improvement in symptoms. He has completed his 6 months course of anti-tubercular therapy and has no recurrence of symptoms.

Conclusion: Musculoskeletal tuberculosis, although uncommon, can manifest with varied features. Reactive arthritis (Poncet's) secondary to tuberculosis or tuberculous tenosynovitisis well described in literature, however, reactive tenosynovitis is very rare phenomenon to occur. High clinical index of suspicion is needed to rule out tuberculosis in such cases.

MIS2023-A-1029

ANCA Associated Vasculitis (AAV) - Multisystem Involvement

HIGHLIGHTS FROM MIDDLE EAST IMMUNOLOGY SUMMIT OF THE GCC AIR

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Introduction: Antineutrophilic cytoplasmic antibody (ANCA) associated vasculitis (AAV) is a small-vessel vasculitis resulting in inflammation of small- and medium-sized blood vessels. Each type of AAV is a rare condition. The incidence is estimated to be between 10 and 20 individuals per million. The prevalence is estimated to be between 200 and 400 individuals per million.

Case report: A 59-year-old male, known to have dyslipidemia, treated hepatitis C infection and recurrent upper respiratory tract infections (URTI). He presented in December 2022 with one month history of cough with hemoptysis. Left ear pain with left eye redness associated with headache and small submandibular swelling noted one day prior to admission. No history of fever, weight loss or skin rash. Patient attended many hospitals and received many antibiotic courses without improvement. He is an ex-smoker. He is allergic to celecoxib and sulfa containing drugs. His regular home medications are aspirin and statin.

On his initial examination, he was afebrile, maintaining oxygen saturation on room air and hemodynamically stable with blood pressure 126/86 mmHg. His systemic examination was normal apart from a red conjunctiva of left eye with no vision abnormalities. Submandibular swelling 1 x 0.5 cm in size, mobile, non-tender. His initial investigations revealed normal electrolytes and renal function. Normal complete blood count and coagulation profile. He had elevated CRP and negative respiratory panel. His urinalysis was positive for proteins and RBCs. Ultrasound neck reported left sided submandibular sialo adenitis. There was no abnormality detected in the CT Head. Computed tomography (CT) chest and CT angiogram showed no evidence of PE, multiple soft tissue nodules in bilateral lungs (neoplasm versus chronic granulomatosis).

Patient was admitted and started on IV antibiotic and in view of the CT chest findings with the suspicion of malignancy versus granulomatous pathology further work up has been sent. TB work up was negative and BAL was negative for granuloma and malignancy. His immunological work up revealed a positive C-ANCA with titer of 155 RU/mL. With the suspicion of AAV a rheumatology input was obtained, their assessment revealed musculoskeletal involvement. Patient was planned for pulse steroids and Nephrology consultation for kidney biopsy and to re-discuss with pulmonology team the need for tissue biopsy.

After the first day of pulse steroids joint pain improved but patient was desaturating overnight and started to require oxygen therapy initially 5 L/min. Decision by rheumatology team was to increase steroid pulse dose to 1 g IV for the 2 consecutive days and to add rituximab. Patient was started on calcium and vitamin D. Nephrology team consulted for kidney biopsy to confirm the clinical diagnosis of AAV. From renal side, renal function was relatively stable Cr 80-90 umol/L, proteinuria 1.5 g/g stable and hematuria improved. Discussion with IR team for CT guided lung biopsy, the consensus was that the risk of pneumothorax is high compared to the desired yield.

Later, patient reported visible hematuria and his creatinine started to rise (peaked up to 200 umol/L) and his proteinuria increased. Kidney biopsy was done and it showed 15 glomeruli with 8 cellular crescents and segmental glomerular fibrinoid necrosis in nine glomeruli consistent with Pauci-immune (c-ANCA+ve) necrotizing and crescentic glomerulonephritis.

In regard to overall patient management, he received IV Methylprednisolone over 3 days 500 mg, 1000 mg, 1000 mg, continued on oral prednisolone initially 60 mg with a tapering regimen. Rituximab 1g, 500 mg and 500 mg each one week apart. Thiopurine methyltransferase Activity was sent and patient was started on Azathioprine initially 100 mg daily which was increased to 150 mg daily in a week time. As the patient was allergic to sulfa containing drugs, he was started on aerosolized pentamidine for PCP prophylaxis.

The question to add cyclophosphamide to his previous management as hybrid regimen was raised as his creatinine was trending up, however it started to platue and improve gradually so cyclophosphamide was not added. Patient clinical and biochemical parameters improved and his repeat CT chest showed improvement and resolution of

lung nodules. He only developed steroid induced DM for which he was managed with insulin and followed up by endocrinology team as outpatient.

Above management was in line with the KDIGO Guidelines for management of AAV, where induction of remission is achieved by glucocorticoids with either cyclophosphamide or rituximab. The RAVE study, a multicentre randomised control non inferiority trial of rituximab as compared with cyclophosphamide for remission induction concluded that rituximab is not inferior to daily cyclophosphamide treatment for induction of remission in severe AAV and may be superior in relapsing disease.

Maintenance therapy with either rituximab or azathioprine and low dose glucocorticoids is recommended after induction of remission. Optimal duration of the maintenance treatment is not known, but should be between 18 months and 4 years.

Conclusion: Associated vasculitis has variable clinical manifestations, early recognition and diagnosis to decrease the risk of life-threatening complications is important. From renal point of view, kidney biopsy is the gold standard for diagnosis, but it should not delay starting immunosuppressive treatment, especially in patients who are rapidly deteriorating. Management of AAV has evolved significantly in the past two decades, with substantial improvements in survival and quality of life. However, more studies are needed especially in regard to duration of maintenance therapy.

MIS2023-A-1026

Trial to Use Icatibant/Lanadelumab in Cases of "IDDM Type 1" Associated with Systemic Angioedema and Urticaria Due to Intrinsic Insulin Allergy

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Case report: Four females of middle age (3 sisters and their niece) with the same history of IDDM Type 1 caused by intrinsic insulin allergy, probable genetically linked. The clinical picture presents as high-grade insulin resistance and idiopathic urticaria resulting in the initial diagnosis of Type 1 – insulin dependent diabetes mellitus. In fact, when started on icatibant the patients recovered their own insulin production – proven by normalized c-peptide levels. Ketones rise when fasting or eating carbs, tests showed normal insulin levels, which is caused by either resistance against insulin versus intrinsic allergy against insulin – deactivating insulin action. The effect of giving insulin resulted in generalized urticaria all over their bodies, itching and lung oedema associated with SOB.

We started icatibant with the 1st patient in May 2022 and she felt much better with the first injection, observing reduced oedema and improved control of her CBG readings between 200-233 mg/dl and while it was up 400 mgdl+ with complication of DKA and admission to the ICU. The first follow up lab test showed a reduction of the HBA1c from >13% to 7.4% after 3 months. And more importantly the patient did not observe any generalized edema nor urticaria. However, when intermittent delay of provision of icatibant delayed her doses (initially twice weekly injections – currently daily injections) this 1st patient started to develop symptoms again like swelling of her face, leg oedema, acute vertigo, neck pain and headache with HbA1c rising to 9.2% again.

In November 2022 this 1st patient was started on lanadelumab as add on but could not be sustained for insurance reasons. Goal would have been to reduce daily injections to twice monthly injection. In November 2022 the 2nd patient was started on catibant - within 6 months – her C-peptide level, initially at zero rose to normal level. By now she cannot be considered DM type 1 anymore.

In March 2023 and April 2023 patients 3 and 4 were started on catibant twice weekly injections with equal improved reduction of HbA1c through improved CBG control via use of their own insulin.

Conclusion: Icatibant was developed for patients with HAE – who have an absence or dysfunction of C1-esterase-inhibitor which leads to the production of bradykinin. The presence of bradykinin may cause symptoms of localized and generalized swelling, inflammation, pain and angioedema. Icatibant inhibits bradykinin from binding at the B2 receptor, thereby treating the symptoms associated with acute attack. Lanadelumab/Takhzyro comes as a further solution as it reduces the dosing to twice monthly.

Currently all 4 patients are either without external insulin doses, or much reduced need for insulin as their own pancreas restarted insulin production. We will need to observe the course and report again next year.

MIS2023-A-1024

Who is the Accused for the New-Onset Myopathy? Ustekinumab vs. Anti-HMG-CoA Reductase Myopathy: A Case Report

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Introduction: Drug-induced myopathy can be defined by a clinical presentation that includes myalgia and muscle weakness, laboratory findings of high creatinine kinase (CK) levels, or the presence of myoglobulin in the clinical context of using some medication known or reported as a cause of myopathy.

Case report: A 50-year-old woman, who had a history of psoriasis and psoriatic arthritis, was admitted because of acute progression of muscle weakness and elevated CK level 9067 U/L after initiation of Ustekinumab. An electromyography (EMG) study showed myopathic potential without muscle irritability. Since the patient was on atorvastatin as well, anti-HMGCR was sent and came to be positive with a titer of 51 U/ml (Ref: negative <20 U/ml). Thyroid stimulating hormone was normal.

The typical clinical findings, along with high CK level and absence of muscle irritability were all consistent with the diagnosis of Drug-induced myositis. Ustekinumab and atorvastatin were both discontinued at this point. Creatinine kinaselevel started to trend down gradually to a normal level without any intervention added over 7 months. The role of positive anti-HMGCR results with high titer has been highlighted recently as useful specific Biomarkers for anti-HMGCR myositis in the setting of a good clinical context (example, presence of muscle weakness, and high CK level in patients with statin use). But still, the course of myositis was self-limited with no immunosuppressive medication added as commonly needed to treat anti-HMGCR myositis. This unique antibody was not detected in self-limited statin myopathy conditions included in some cohort studies previously.

Conclusion: In the end, we conclude that Ustekinumab was the likely trigger of myositis in our case. There are tow cases reported previously of myositis associated to Ustekinumab use. This potential adverse event should be taken into account when we follow our patients who are on Ustekinumab therapy. The knowledge is lacking behind the exact underlying pathophysiology.

MIS2023-A-1021

Inferior Vena Cava Thrombosis in Patient with Bechet's from Nairobi, Kenya

HIGHLIGHTS FROM MIDDLE EAST IMMUNOLOGY SUMMIT OF THE GCC AIR

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Introduction: Thrombosis in Behcet's occurs in about 8-12% whereas vena cava thrombosis occurs in about 4 -8.2%. Thrombosis include: deep vein thrombosis, superficial thrombophlebitis, vena cava thrombosis, pulmonary thromboembolism, cerebral venous thrombosis (CVT), intracardiac thrombosis, Budd–Chiari syndrome (7.5%) and Renal vein thrombosis. Venous thrombosis is more frequent than arterial thrombosis, and multiple thrombosis is frequent.

Management of thrombosis includes: glucocorticoids and immunosuppressive such as azathioprine, cyclophosphamide or cyclosporine-A are recommended. Deep vein thrombosis is thought to result from inflammation of the vessel wall rather than hypercoagulability. For deep vein thrombosis, immunosuppressants alone are recommended. For other sites, anticoagulants have been used (caution: need to ensure no pulmonary aneurysm)

Case report: A description of patient, 51-year-old female, hairdresser. Presented with shortness of breath, fatigue, recurrent and persistent painful oro-genital ulcers, and chest pain despite treatment with colchicine, azathioprine and prednisolone.

Tests done were negative or normal. Erythrocyte sedimentation rate of 18 mm/hour, normal total blood count and liver function tests, folic acid and ferritin. The HIV/HCV/HBSAG: negative, VDRL/ HSV serologies: negative, ANA, ENA, DSDNA negative, normal complement c3 and c4, negative pathergy test. Malignancy screen and antiphospholipid antibodies were negative.

She was on colchicine, azathioprine with reduction in ulcers. She had persistent dyspnea, low oxygen saturation 84% on room air, CT pulmonary angiogram no pulmonary thromboembolism but was noted to have a filling defect in the IVC in keeping with IVC thrombosis. She was initiated on anticoagulant in view of being on golimumab and concern about pulmonary microembolism with resultant dyspnea and hypotension. Her medication was changed to infliximab at 5 mg/kg and administered as per schedule.

She had resolution of dyspnea and the orogenital ulcers and continues on her medication that includes infliximab, colchicine and prednisolone 5 mgs daily .

Conclusion: We described a 51 year old female with inferior vena cava thrombosis who was successfully treated with infliximab.

MIS2023-A-1019
Idiopathic Orbital Inflammatory Syndrome- A Case Series

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Introduction: Idiopathic orbital inflammatory disease is an inflammatory eye disease causing non granulomatous changes on histopathology, frequently mistaken for pre-septal or orbital cellulitis. While it is commonly encountered among adult age group it is quite rare among pediatrics.

Case report: To report a case series of 2 patients who presented with bilateral eye swelling. An 11-year-old previously healthy girl and a 12-year-old girl known to have type 1 diabetes mellitus were both evaluated for bilateral eye swelling. On examination, conjunctivitis and restriction of the eye movement were evident in both

patients. Laboratory work up showed negative inflammatory markers in the first patient while they were elevated in the second patient, furthermore both patients had negative infectious work up including tuberculosis, syphilis and HIV, and ACE levels. Both patients were treated initially with antibiotics. Moreover; MRI orbit was ordered illustrating bilateral myositis in the first patient and diffuse lacrimal gland enlargement in the second patient raising the suspicion of idiopathic orbital inflammatory disease, however further investigations weer required to establish a diagnosis for the second patient hence she underwent a biopsy of the lacrimal gland where the histopathology report showed severe active chronic inflammation associated with stromal fibrosis and no granulomas. Treatment with corticosteroids was commenced for both patients resulting in significant clinical improvement and further confirming the diagnosis, however due to the ongoing inflammation the second patient was started on Methotrexate. Both patients have regular follow up with both rheumatology and ophthalmology services.

Conclusion: Idiopathic orbital inflammatory disease is a rare entity among pediatric age group. Yet, an early diagnosis through the joint care with the ophthalmology team and prompt treatment are vital to prevent complications such as retinal and extraocular muscle damage.

MIS2023-A-1041
A Rare Case of Dominantly Inherited Periodic Fever

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Introduction: Autoinflammatory diseases, genetic disorders impacting the innate immune system. Among these, Tumor necrosis factor autoinflammatory diseases are a genetically inherited group of disorders that affect the innate immune system. Dominantly inherited periodic fever syndromes are poorly understood among physicians, especially in our region. They have been described in Caucasians. Tumor necrosis factor receptor-associated periodic fever (TRAPS) is a very rare disease that falls under the category of dominantly inherited periodic fever, with an estimated prevalence of one per million. It was first described in 1982 as "Familial Hibernian Fever" and was later renamed to TRAPS after genetic basis were revealed. The disease is caused by mutation in TNF superfamily receptor 1A (TNFRSF1A). The TRAPS is characterized by attacks of fever lasting around 5-25 days and associated with other non-specific symptoms as abdominal pain, joint pain, rash, fatigue, and so on. Most severe complication is AA-type amyloidosis. Diagnosis relies on high clinical suspicion supported by genetic findings.

Case report: A 12-year-old Emirati boy presented with a history of recurrent, prolonged fever, starting at 1 year and 6 months of age. He experienced multiple fever episodes per year, each lasting 1-2 weeks, accompanied by severe abdominal pain. His fever reached 39-40°C and persisted. He underwent repeated hospitalizations across different medical centers, receiving intravenous antibiotics. Laboratory tests revealed elevated inflammatory markers, yet no identifiable infection source. A trial of colchicine proved ineffective. At age 9 years, genetic testing detected a heterozygous nucleotide exchange in exon 3 of the TNFRSF1A gene, resulting in an amino acid substitution (p.Thr79Met or T50M), associated with TRAPS. Treatment commenced with Canakinumab monthly injections.

The TRAPS is a rare syndrome that is occasionally seen around the world. It is associated with variable mutations in the TNFRSF1A gene on chromosome 12. Primarily within exon 2 and 4 and are due to missense substitutions that interrupt important cysteine-cysteine disulfide bonds.

The TRAPS remains an enigmatic autoinflammatory disease, particularly in regions with limited prevalence.

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Timely diagnosis and appropriate treatments are imperative to prevent severe complications. Further research is necessary to elucidate the relationship between initial clinical presentation, disease progression, and complications. **Conclusion:** TRAPS is a rare syndrome that is occasionally seen around the world. It is associated with variable mutations in the TNFRSF1A gene on chromosome 12. Primarily within exon 2 and 4 and are due to missense substitutions that interrupt important cysteine-cysteine disulfide bonds. This case report describes a 12-year-old Emirati boy diagnosed with TRAPS. The TRAPS remains an enigmatic autoinflammatory disease, particularly in regions with limited prevalence. Timely diagnosis and appropriate treatments are imperative to prevent severe complications. Further research is necessary to elucidate the relationship between initial clinical presentation, disease progression, and complications.

MIS2023-A-1009

Dermatological Manifestation of COVID-19: A Rare Presentation of Immunoglobulin a Vasculitis in a Hemophilia A patient

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Introduction: COVID-19 has affected millions worldwide, primarily presenting with respiratory symptoms. Dermatological manifestations are rare but have been reported. We present a rare case with hemophilia A presenting with immunoglobulin A vasculitis (IgAV), also known as Henoch-Schönlein purpura, as a complication of COVID-19.

Case report: A 23-year-old man diagnosed previously with hemophilia A presented with a one-week history of skin rash and abdominal pain. The abdominal pain was colicky, non-radiating, and worsened with eating. It was associated with fatigue, bodyache, and decreased oral intake. He did not respiratory symptoms of COVID-18 infection. Skin rash initially in the lower limbs, spread to the upper extremities. There was no associated itching or pain. He also reported watery diarrhea with no blood or mucus. A review of other symptoms was unremarkable. On physical examination revealed a purpuric and petechial rash on the lower and upper extremities. Abdominal examination was unremarkable, with no organomegaly.

Laboratory results revealed a slight elevation of CRP (8.3 mg/L, NR 0 - 5), with normal urea and electrolytes, complete blood count, liver function tests, and coagulation profile including Factor VIII activity (3.90%). Immunological tests showed negative ANA, cANCA, pANCA, rheumatoid factor, and cryoglobulin. CT abdomen with contrast showed no signs of vasculitis. A skin biopsy strongly indicated IgA vasculitis. On the 4th day of admission, nasopharyngeal swab was positive for COVID-19 infection.

The patient was started on intravenous methylprednisolone (40mg daily). COVID-19 was managed symptomatically. Hematology advised the continuation of anti-hemophilic factor VIII (1500 units three times per week) due to low disease activity. The patient was discharged on a tapering dose of prednisolone (40mg daily) with a follow-up plan in the rheumatology clinic.

A literature review revealed only a few case reports about the association between IgAV and hemophilia patients, mostly in pediatric patients. To our knowledge, this is one of the few reported cases of IgAV associated with COVID-19 in an adult with underlying hemophilia A.

Conclusion: Timely recognition and management of dermatological complications, such as IgAV, in COVID-19 patients who have hemophilia can contribute to improved clinical outcomes. Further research is warranted to better

understand the pathophysiology and prevalence of IgAV in the context of COVID- 19, as well as the long-term implications for affected individuals.

MIS2023-A-1005

A Confusion With a Positive Antinuclear Antibody Test

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Introduction: Bechet's disease is a complex, chronic inflammatory disorder that affects multiple systems in a relapsing-remitting status. The exact etiology remains unknown, but the hallmark of the disease is arterial/venous vasculitis manifestation. It is diagnosed mainly clinically, which adds more challenges in management and treatment. The diverse range of symptoms in Bechet's disease underscores its diagnostic complexity. We are reporting two challenging cases that fulfill the International classification of Bechet disease (ICBD) 2006 criteria with potential involvement of the cardiovascular and central nervous systems respectively.

Case report: Case 1: A 43-year-old Emirati male diagnosed with Bechet disease presented to the ER with chest pain radiating to the left shoulder. Electrocardiogram (ECG) revealed an inferolateral myocardial infarction (MI). Cardiac catheterization identified an ectatic lesion in right coronary artery (RCA) and left coronary artery , which was managed through stenting. Despite successful Percutaneous Cardiac Intervention (PCI), the patient continued to experience recurring atypical chest pain without ECG findings nor elevation of cardiac markers for a year. A subsequent PCI documented occlusion of RCA and the presence of multiple coronary luminal irregularities of the LAD and the RCA, suggesting active vasculitis. The patient commenced on rituximab infusion followed by azathioprine and maintained at remission.

Case 2: A 23-year-old Sudanese male, presented to the ER with a one-week history of severe headache, fever, and blurred vision. Prior to this presentation, the patient declared to have four episodes of red painful eyes, multiple painful mouth ulcers, and two attacks of genital ulcers in the past year. Brain MRA was negative, however, Magnetic Resonance venography (MRV) showed a large clot in the right sigmoid sinus and internal jugular vein suggestive of sinus thrombosis. Antibiotics and anticoagulants were initiated, but there was no improvement. Based on the above clinical manifestation and positive neuro-radiological findings, a diagnosis of acute neurobehect disease was established. Pulse steroids regime followed by infliximab infusions resulted in significant improvement.

Conclusion: Vascular manifestation of Behect disease can be devastating with a fatal outcome if not diagnosed early with prompt management. Additional research is required to enhance our understanding of the underlying pathophysiology and optimize management strategies for Bechet's disease.

MIS2023-A-1007

Ulcerative Colitis Masquerading as Polymyalgia Rheumatica

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Introduction: This case report discusses the diagnostic challenge posed by a 64-year-old female patient with a complex medical history, including ulcerative colitis, who presented with acute onset generalized pain, particularly

in the bilateral shoulders and hips. The patient's clinical presentation initially suggested polymyalgia rheumatica (PMR), but the diagnostic process revealed an unexpected twist.

Polymyalgia rheumatica is an inflammatory condition primarily affecting older adults. It typically presents with proximal muscle pain and stiffness. However, the clinical presentation can overlap with other conditions, making diagnosis challenging, especially in patients with multiple comorbidities.

Since there is no definitive test to diagnose PMR, the diagnostic process relies on the ACR/EULAR 2012 provisional classification criteria. These criteria added the value of using ultrasound in the assessment of findings, which may include subdeltoid bursitis, biceps tenosynovitis, glenohumeral synovitis, synovitis, or trochanteric bursitis.

Case report: The patient is a 64-year-old female with a history of diabetes mellitus type 2, ischemic heart disease, hyperlipidemia, ulcerative colitis (pancolitis), stage 3 chronic kidney disease, osteoporosis, and functional dyspepsia. She presented to the rheumatology clinic with the following complaints, acute onset generalized pain with a focus on bilateral shoulder and hip pain lasting for 5 days, inability to lift her arms above shoulder level, difficulty walking due to bilateral hip pain, morning stiffness lasting more than 30 minutes and fatigue. Review of symptoms revealed patient has ongoing abdominal pain associated with intermittent bloody stool for the past 6 months. Musculoskeletal examination of the bilateral shoulders revealed tenderness along the joint line, a restriction in the arm's range of motion and bilateral hip tenderness at the trochanteric area.

Signs of inflammation were given by raised ESR 110 mm/hour and CRP 65 mg/l, HB 11 mg/dl , GFR 48 and calprotectin >2100 ug/ml. While other basic blood tests were normal. Autoimmune diseases were ruled out by negative antinuclear antibody, rheumatoid factor, and anticyclic citrullinated peptide antibody. Bilateral shoulder x-ray shows osteoarthritic changes and ultrasound findings demonstrates subacromial subdeltoid bursitis. The patient's complex medical history, including refractory ulcerative colitis (sulfasalazine) and polymyalgia rheumatica (PMR)-like symptoms, posed a significant therapeutic challenge. After a comprehensive evaluation, a novel approach was considered.

The patient was initiated on ustekinumab, a monoclonal antibody targeting interleukin (IL)-12 and IL-23, primarily as a treatment for refractory ulcerative colitis. Remarkably, over the course of one year of ustekinumab therapy, the patient experienced significant improvement in her overall clinical condition. Her PMR-like symptoms, including acute bilateral shoulder and hip pain, morning stiffness, and fatigue, notably subsided.

This favorable response to ustekinumab not only led to the successful management of her ulcerative colitis but also provided relief from the debilitating symptoms of polymyalgia rheumatica. The sustained improvement in her condition emphasizes the potential of ustekinumab as a therapeutic option for patients with concurrent inflammatory conditions, such as ulcerative colitis and PMR-like symptoms.

Conclusion: The patient's response to treatment underscores the importance of personalized and innovative approaches in managing complex cases with overlapping symptoms and comorbidities.

MIS2023-A-1006

Systemic Lupus Erythematosus Presenting as Evans Syndrome

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Introduction: Evans syndrome (ES) is a rare syndrome, it is characterized by the presence of autoimmune hemolytic

anemia, immune thrombocytopenia, or immune neutropenia of unknown etiology. It occurs mainly in children, usually due to primary immunodeficiencies or autoimmune lymphoproliferative syndrome. As the hematological abnormalities are an important diagnostic criterion for systemic lupus erythematosus (SLE), we present a patient diagnosed with ES with underlying SLE on initial presentation.

Case report: A 47-year-old Saudi female, with a past medical background of sickle cell trait, hypothyroidism on levothyroxine, pancytopenia followed with a hematologist and managed her as a case of myelodysplasia for 6 years, refractory to recurrent blood, and platelets transfusion, bone marrow biopsy was done 3 times in the past showed hypercellular reactive bone marrow with trilineage hematopoiesis no evidence of myelodysplasia . She recently encountered a COVID-19 infection one month before presentation. The patient presented to our hospital with a long history of generalized weakness, fatigue, dizziness, and exertional dyspnea. Her symptoms gradually worsened over time that she sever fatigue and dizziness on standing. She reported easily bruising on her extremities, recurrent gum bleeding after brushing her teeth, epistaxis, and menorrhagia which resulted in recurrent iron deficiency anemia.

Workup upon this presentation showed hypochromic microcytic anemia with hemoglobin 5.1 g/dL, total leukocyte counts 1.9 X103/uL [4-10], red blood cells 2.32 [3.8-4.8], and platelets 2 X103/uL [150-430], with neutropenia (Neutrophil count 0.96x10^3/uL) [1.5-6] and lymphopenia (Lymphocyte count 0.46 X10^3/uL) [1.3-2.9] in the differential count. Positive direct Coombs test and peripheral blood film showed moderate leukopenia, slight hypochromic microcytosis, slight teardrop cells, target cells, and thrombocytopenia. Her reticulocyte count was 6.29% [3-6], and her lactate dehydrogenase (LDH) 382 IU/mL [135-225]. Further labs showed low C3 (0.57 g/L) normal C4 (0.18 g/L), ANA 1:320 nucleolar, and anti-DNA ABs (57 IU/ml), anti SSB (LA): Positive. Bone Marrow aspiration & trephine biopsy: Reactive cellular marrow and intact trilineage hematopoiesis with no evidence of dysplasia or infiltration.

Conclusion: Given the patient's clinical presentation with the above laboratory findings, we diagnosed the patient as a case of Evan syndrome secondary to SLE, as the patient has hemolytic anemia, immune thrombocytopenia, lymphopenia and neutropenia. She received IV pulse steroid for 3 days, then shifted to oral tapering down steroid, Intravenous immune globulin (IVIG) for 5 days, and eltrombopag olamine. She showed a marked improvement clinically, as well as continued improvement in her anemia and thrombocytopenia after starting her on treatment.

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Unusual Presentation of Pediatric Polyarteritis Nodosa: Case Report

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Introduction: Polyarteritis nodosa (PAN) is a rare systemic necrotizing vasculitis of small and medium sized arteries that leads to thrombus, infractions and aneurysms in various organs. PAN is most common in males between 40 and 60 year and rare in childhood. The central nervous system is much less commonly involved than peripheral nervous system. We present a case of juvenile PAN with cerebral aneurysm as initial presentation which was successfully treated with embolization. High dose of steroids and cyclophosphamide was given as induction therapy with improvement.

Case report: A 6-year-old male patient presented with fever, weight loss, and livedo reticularis, then he developed

headache with projectile vomiting. Computed tomography (CT) brain showed subarachnoid hemorrhage. The CT angiography showed small saccular aneurysm along course of right anterior temporal branch of right middle cerebral artery. Two small mm aneurysm along distal course of right anterior cerebral artery and occiptopariteal branch of right posterior cerebral artery. Right anterior temporal and right superior cerebellar artery ruptured aneurysm for urgent embolization. The patient was referred to our department. Infectious causes for cerebral aneurysm were excluded. Pan cultures was negative. Echocardiography was normal. Serum titre for antinuclear antibody, ds-DNA, P-ANCA, C-ANCA were all-negative. The patient was diagnosed as PAN based on diagnostic criteria for childhood PAN.

High dose of steroids and cyclophosphamide were given with improvement. The patient was maintained on low dose of steroids and 100 mg of Azathioprine for 6 years. In 2021 the patient stopped his treatment after which he presented with colicky abdominal pain, severe progressive vomiting, and bloody diarrhea. The CT abdomen showed small bowel obstruction with necrotizing enter colitis. Picture suggestive of mesenteric vascular occlusion. Resection and anastomosis was done. The CT angiography of abdominal aorta and mesenteric vessels showed patent abdominal aorta, its main branches (celiac, superior and inferior mesenteric as well as renal arteries. Patent portal vein and its branches. No evidence of aneurysm or dissection. The patient was kept on steroids (0.5 mg/kg) and mycophenolate mofetyl (2gram).

In 2023 the patient started to complain of acute diminution of vision. Urgent magnetic resonance imaging (MRI), magnetic resonance venography (MRV), and magnetic resonance angiography (MRA) were done. The MRI brain reported early occipital subacute hematoma. Right cerebellar areas of encephalomalacia. The MRV and MRA showed notable tuft of serpiginous arteries arising from the right middle cerebral artery, mildly dilated cortical veins is noted at the overlying fossa reaching to the superior sagittal sinus, attenuated left vertebral artery with dominant right one with normal MRV of the brain. Pulse corticosteroid was given for 3days then 0.5 mg/ kg of oral steroids. Re-induction with cyclophosamide was started. The patient was treated by embolization, steroids and cyclophosphamide

Conclusion: We reported a rare case of CNS involvement in a paediatric case with PAN who was treated successful with corticosteroids, cyclophosphamide and embolization.

Category: Clinical Research

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Effects of post- COVID-19 Vaccination on Circulating Cytokine Profile

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Backgrounf: The worldwide emergence of the COVID-19 pandemic led to the expeditious development and implementation of vaccinations to prevent infection and contain the spread of disease Understanding the dynamics of immune responses following vaccination is critical for optimizing vaccine strategies in future pandemics. In this retrospective longitudinal investigation we analyzed the effect of COVID19 vaccination on circulating cytokine profile in individuals living in Saudi Arabia.

Methods: A total of 318 Saudi subjects (59.7% females), comprising individuals of varying ages from 12-60 years,

received COVID-19 vaccines as per the national vaccination program. Anthropometric data and fasting blood samples were collected at specific time points pre-and post-vaccination. Information on dates of vaccination, and whether or not infected with COVID during the study period were collected. For this study, the samples from 84 subjects were used for a comprehensive 18-parameter cytokine profiling analysis using state-of-the-art techniques. The participants were stratified into 2 groups based on the interval between the final vaccine dose and follow-up visits.

Preliminary findings indicate that circulating cytokine profiles are importantly impacted by vaccination. Notably, when shorter (≤4 months) and longer (≥5 months) intervals between the final dose and follow-up were compared, significant differences in cytokine profiles were observed, as follows: interleukin-1β (6.5 (0.8 - 12.3) pg/ml versus 0.0 (-1.3 - 4.3) pg/ml, p=0.024), interleukin-7 (6.2 (1.5 - 9.5) pg/ml versus (vs.) 0.8 (-2.4 - 4.3) pg/ml, p=0.001), tumor necrosis factor-alpha (1.0 (-2.4 - 10.5) pg/ml vs. -0.8 (-5.7 - 4.5) pg/ml, p=0.028), and monocyte chemoattractant protein-1 (95.1 [-25.7 - 259.7] pg/ml vs. -6.5 [-83.8 - 84.1] pg/ml, p=0.019).

Results: Preliminary findings indicate that circulating cytokine profiles are importantly impacted by vaccination. Notably, when shorter (≤4 months) and longer (≥5 months) intervals between the final dose and follow-up were compared, significant differences in cytokine profiles were observed, as follows: interleukin-1β (6.5 (0.8 - 12.3) pg/ml vs. 0.0 (-1.3 - 4.3) pg/ml, p=0.024), interleukin-7 (6.2 (1.5 - 9.5) pg/ml vs 0.8 (-2.4 - 4.3) pg/ml, p=0.001), tumor necrosis factor-alpha (1.0 (- 2.4 - 10.5) pg/ml vs. -0.8 (-5.7 - 4.5) pg/ml, p=0.028), and monocyte chemoattractant protein-1 (95.1 [- 25.7 - 259.7] pg/ml vs. -6.5 [-83.8 - 84.1] pg/ml, p=0.019).

Conclusion: This longitudinal study sheds light on nature of the immunologic response induced by COVID-19 vaccination and indicates that vaccine-induced cytokine production wanes in-time after the last dose of vaccine Further research is required to investigate the stability of these cytokine profiles over time and to determine their relationship with vaccine efficacy and long-term immunity.

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Explore the Prevalence of Various Food and Inhalation Allergen's Specific IgE Antibody Positivity in Residents of Northern Emirates of UAE: EUROLINE Food Middle East 2 and Inhalation Gulf (IgE) Test Based Pilot Study

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Background: The aim of the present study was to explore the prevalence of various food and inhalation allergen's allergen-specific IgE antibody positivity in residents of Northern Emirates of UAE using the Immunoblot technique. The term allergy is the body's increased ability to react to a foreign substance and today the term allergy means an oversensitivity to foreign substances which are normally harmless. Alongside any genetic predisposition, numerous non-genetic factors also play a role, such as exposure to the allergen, nutritional condition, existing chronic diseases, and acute viral infections. A food allergy (FA) is a set of immunological responses that appears to happen when a person shows a particular interaction with some food proteins. FA is classified into three types based on the immune-mediated response: immunoglobulin (IgE)-mediated and non-IgE mediated or a combination of both. Inhaled allergens are divided into two groups including outdoor such as pollens and molds, and indoor allergens such as house dust mites, animal dander, and molds.

Methods: A A total of 255 patients blood samples were received from various hospitals and clinic from the Northern Emirates region to Thumbay Laboratory, Thumbay University hospital, Ajman to perform various allergy tests including food and inhalation allergy during the study period of 02 years (Jan 2021 to Dec 2022). Using EUROLINE immuno blotting method, 47 different food allergens impregnated latex strips were used to perform the food allergy test on the 188 patients' serum samples, 46 serum samples were used to do the inhalation allergy and 21 samples were also used to do both tests. The patients' demographic details and their age groups (≤1 year, 1–10-years, 11–20-years, 21-30-years, 31-40-years, 41-50-years and ≥ 60-years-old) were also included.

Results: Out of 255 samples, 63.2% (n=161) samples were positive to allergic tests and 36.8% (n=94) were negative. Among the positive samples, 119 (73.9%) samples were only positive to various food allergens, 30 (18.6%) were only positive to inhalation allergens and 12 (7.5%) samples were positive to both food and inhalation allergens. Among the 161 positive cases, 89 (55.3%) were male patients and 72 (44.7%) female patients. In that 161 positive cases, 92 (57.14%) were from Arab ethnicity and 69 (42.86%) were non-Arab ethnicity. Among the positive patients, 42.85% (n=69) were belong to 21- 40 years-old patients and 20.49% (n=33) were belongs to 1-10-years-old children.

Most of the children under the age group of ≤10 years-old were most predominantly allergic to cow milk UTH with the IgE concentration of 3.5 ≤ - <50.0 kU/l and the remaining age groups (1–10-years, 11–20- years, 21-30-years, 31-40-years, 41-50-years and ≥ 60-years-old) were most commonly allergic to shell fish and fishmix, and then cow milk, chicken and other allergens. Like this, all the patients were predominantly allergic to dust mites; dermatophagoides pteronyssinus and dermatophagoides farina.

Conclusion: The study concludes that the most prevalent food allergens were shellfish mix 4, cow milk (UTH), fish mix, Pistachio, chickpea, fruit mix, peanut, sesame, garlic, almond and orange. Similarly, in the inhalation allergy