Clinical features of systemic sclerosis

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ABSTRACT

Objectives: This descriptive study was carried out to determine the clinical and epidemiological pattern of systemic sclerosis in Iraqi patients who prove to have the disease.

Methods: Between March 1997 and December 1999, 75 patients, 67 females and 8 males, were studied at 2 teaching hospitals in Baghdad, Iraq. The diagnosis of the disease was based on clinical findings, biochemical, serological, and pulmonary function tests.

Results: All the patients had Raynaud's phenomenon, 72 (96%) had arthralgia, 65 (87%) had dysphagia, 62 (83%) had atrophic changes or hypo/hyperpigmentation, 43 (58%) had telangiectasia, and 3 (4%) had subcutaneous calcification. Of the patients studied, 2 patients (3%) had hypertension, one had severe hypertension. Twenty-three patients (31%) had anemia, 58 (77%) had elevated erythrocyte sedimentation rate and 7 patients (9%) with renal impairment. Rheumatoid factor was positive in 17 patients (23%) and antinuclear antibody in 50 (67%). Abnormal chest x-ray was reported in 50 patients (67%). Pulmonary function tests showed restrictive pattern in 53 (71%) patients and obstructive pattern in 3 (4%). Electrocardiogram changes were seen in 3 (4%) of the patients. Three juvenile cases were reported, and familial tendency was noted in one patient. The results were compared with other studies and there were interesting differences and similarities.

Conclusion: Systemic sclerosis is a rare disease in Iraq, nevertheless sporadic cases at a peak age of 20-40 years were reported. Generally the clinical picture was comparable to other studies, however Raynaud's phenomenon is more prevalent in our patients and antinuclear antibody is less common.

Keywords: Systemic sclerosis, epidemiology.

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Systemic sclerosis is an uncommon multi-systemic disease of unknown etiology. It is characterized by extensive inflammation and fibrosis of the skin and viscera, associated with proliferative vascular and microvascular lesions.[1] The incidence varies between 2.3 to 12 cases per million of the population.[2] It may present at any age with a peak incidence in the 3rd and 4th decades of life, and is rarely recognized in children.[3] There is an occupational preference especially among coal and gold miners.[4] There is no significant racial differences in incidence, however the prognosis seems to be worse in Blacks than in Caucasians.[5] Although systemic sclerosis is an uncommon disease, sporadic cases are reported in Iraq. This study was carried out to highlight the clinical and epidemiological features of this disease among Iraqi patients at 2 teaching hospitals.

Methods. Between March 1997 and December 1999, a consecutive 75 patients were seen at 2 teaching hospitals. The female to male ratio was 8.4:1. Eight patients (one of them male) were

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Kurdish and the rest were Arabs. The patients were selected on inclusion and exclusion criteria.

Inclusion criteria. Any patient who has fulfilled American Rheumatism Association (ARA) criteria.[6]

Exclusion criteria. Any patient with CREST syndrome alone or mixed connective tissue disease and overlap syndrome.

A detailed medical history was taken from all patients including sex, age, residency, occupation, ethnic group, social status, duration of illness and complications. Also, a full medical examination was carried out on every patient including mouth-opening distance and chest expansion in centimeters at the 4th intercostal space. Duration of the disease was taken from the onset of symptoms until the time of admission of the patients to the study. All patients performed the following investigations (with limitation due to the current status of the country): complete blood picture (CBP), erythrocyte sedimentation rate (ESR), blood urea, serum creatinine, serum alkaline phosphatase and general urine examination, latex fixation test for rheumatoid factor and antinuclear antibody (ANA), chest x-ray, barium swallow and x-ray of both hands, electrocardiogram (ECG), pulmonary function test including forced vital capacity (FVC), forced expiratory volume in the first second (FEV1) and peak expiratory flow rate (PEFR). Results were compared with the normal predicted values according to the World Health Organization classification criteria. Unfortunately diffusing capacity and spinal computerized tomography (CT) scan are not available at the 2 institutes.

Results. Sixty-seven patients were female and 8 were male with a female to male ratio of 8.4:1, and the peak age incidence was at 20 to 39 years (Table 1). There was a positive family history in one case in which the mother and the daughter were affected. The disease's prominent symptoms are shown in Table 2. Moderate hypertension was seen in 2 patients (3%) and severe hypertension in one patient. Anemia presented in 23 patients (31%) and the number of patients having a high white blood cell (WBC) count was 7 (9%). Fifty-eight patients (77%) showed elevated ESR, only 5 of them (7%) had an ESR level more than 100 mmHg. Surprisingly, rheumatoid factor was positive in those patients although none of them fulfilled the ARA criteria of rheumatoid disease. Serum alkaline phosphatase was elevated beyond the normal level in 15 patients (20%). Antinuclear antibodies by the immunofluorescence technique was positive in 50 patients (67%), ANA titre was 1:160 in 19 patients, 1:320 in 47, 1:640 in 8 and only one patient with a tire of 1:1280. Rheumatoid factor was positive in 17 patients (23%). We were unable to perform double stranded deoxyribonucleic acid (DNA) and anti RNP due to the current situation of the country. General urine examination showed albuminurea and granular casts in 3 (4%) patients. Seven patients (9%) showed elevated blood urea, 3 of them (4%) showing albumin urea and urinary sediments. One of our patients died from renal failure. Fifty patients (67%) showed features of systemic sclerosis in their chest x-ray. Forty-six out of 50 had basal reticulonodular shadowing, 6 of them with extensive basal honeycomb shadowing. The other 4 patients had reticulonodular shadowing at the upper and middle lobes. Two patients presented with aspiration pneumonia during the period of the study. Forty-seven patients (63%) presented with typical systemic sclerotic changes in their barium swallow. Regarding the pulmonary function tests, 19 patients (25%) had normal test results while the rest had abnormal results; 53 patients (71%) had a restrictive pattern and only 3 (4%) of them had an obstructive pattern as well. Neurological involvement (trigeminal neuralgia) was found in 3 patients (4%) and carpal tunnel syndrome in 6 patients (8%). Only 2 patients (3%) showed abnormal ECG changes of right bundle branch block.

Table 1 - Age and sex distribution of cases.

Age Group Years	Male Number (%)	Female Number (%)	Total Number (%)
20	1 (1)	5 (7)	6 (8)
20-29	4 (5)	17 (23)	21 (28)
30-39	1 (1)	19 (25)	20 (27)
40-49	1 (1)	13 (17)	14 (19)
50-59	1 (1)	10 (13)	11 (15)
60 & over	- -	3 (4)	3 (4)
Total	**8 (9)**	**67 (89)**	**75 (101)**

Table 2 - Signs and symptoms presentated in our study and a comparison with other studies.

Signs & Symptoms	Our study Patients No (%)	Other studies
Raynaud's phenomenon	75 (100)	75%[7]
Arthralgia	72 (96)	-
Dysphagia	65 (87)	90%[8]
Hypo/Hyperpigentation	62 (83)	-
Atrophic chances	62 (83)	-
Telangiectasia	43 (58)	32%[9]
Subcutaneous calcifications	3 (4)	7%[9]

Discussion. Although systemic sclerosis is believed to be a rare disease in comparison with other collagen diseases in rheumatology practice in Iraq, we were able to collect 75 patients in less than a 3 year period at 2 teaching hospitals. The patients' ages ranged between 14 and 70 years with a peak incidence in the 3rd and 4th decades. This is different from other studies that showed a peak incidence in the 4th and 5th decades of life.[1] The disease was reported rarely to affect children,[3] however, in our study 3 cases of juvenile type were registered. Females constituted 89% of our patients compared to 73% in other studies.[2] No significant observations were detected regarding the occupation or socio-economic status. Although some familial cases were reported by other studies, we found only one case of a mother and her daughter being affected.[10] However, when Tuffanelli and Winkelmann[11] reviewed the literature, he studied 727 cases of progressive systemic sclerosis and found no familial occurrence. The signs and symptoms of systemic sclerosis differ from our study in comparison to the different other studies published in the literature (Table 2). Raynaud's phenomena and telangiectasia were more frequent in this study. Three patients (4%) were hypertensive, 2 with moderate hypertension and renal impairment. LeRoy et al study showed hypertension in 10% of their patients,[12] while Helfrich et al studied 140 patients and found that 15 out of 131 had normotensive renal failure.[13] Applying the criteria of renal involvement (azotemia, proteinurea and hypertension), 7 patients (9%) had renal involvement which was much lower than 35% in other reports.[12]

Though the majority of our patients had dysphagia, only 23 (31%) patients were anemic, 6 of them showed hemoglobin levels less than 10 g/dl. On the other hand, all patients with anemia were found to have dysphagia as well. Frayha et al[9] explained the anemia with systemic sclerosis to be related to dysphagia. Seven patients (9%) had leukocytosis, which is comparable to other studies.[14] Fifty patients (67%) showed positive ANA however Bernstein[15] reported a prevalence of more than 90% positive ANA with systemic sclerosis. Rheumatoid factor was positive in 22% of our patients compared to 25% in other studies.[16] Pulmonary involvement was common in our patients, 67% had positive chest x-ray findings. Pulmonary function tests revealed a restrictive pattern in 53 (71%) patients and only 3 (4%) had an obstructive pattern. Steen et al reported 71% of their patients to have a restrictive pattern and 12% had an obstructive pattern.[17] Although all our patients complained of dysphagia, only 63% of our patients have changes correlated with the severity of the disease on Barium swallow in comparison to 75% as reported by Lock et al.[18] Only 2 patients (3%) showed ECG changes compared to 7% in other studies,[19] and 42% had cardiac rhythm abnormalities.[20] Neurological manifestations (Trigeminal neuralgia) were seen in 3 patients (4%) only, compared to 6% in other studies.[21]

In conclusion, the findings of this study show that systemic sclerosis is a not rare disease at 2 Iraqi teaching hospitals. This disease presented sporadically at a peak age of 20-39 years. Raynaud's phenomenon and telangiectasia is a more frequent presentation in our patients than in other studies and ANA is less common. There was one familial occurrence and one mortality during the period of the study.

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