Maxillary sinus nodular fasciitis

Abdulmalik S. Alsaied, MD, *Mohammed A. Lotfy,* MD.

ABSTRACT

تعد التهاب اللفافة العقيدي حالة نادرة والتهاب حميد ولكن قد تشخص خطأً كورم خبيث. نستعرض هنا حالة نادرة لالتهاب اللفافة العقيدي الناتج من الجيب الفكي لدى طفل يبلغ من العمر عامان. وبمراجعة المنشورات الطبية الإنجليزية وجدنا 3 حالات نشرت كالتهاب اللفافة العقيدي في الجيوب الأنفية وبطرق علاجية مختلفة لكل حالة.

Nodular fasciitis is a rare and benign inflammatory condition; however, it can be misdiagnosed as a malignant lesion. We report a unique case of nodular fasciitis arising from the maxillary sinus in a 2-year-old child. Our English literature review (PubMed search), revealed a total of 3 cases published as nodular fasciitis in the para-nasal sinuses, each with a different management approach.

Saudi Med J 2014; Vol. 35 (1): 88-92

From the Department of Otorhinolaryngology, College of Medicine, University of Dammam, King Fahd Hospital of the University, Dammam, Kingdom of Saudi Arabia.

Received 21st July 2013. Accepted 25th November 2013.

Address correspondence and reprint request to: Dr. Abdulmalik S. Alsaied, Senior Registrar, Department of Otorhinolaryngology, College of Medicine, University of Dammam, King Fahd Hospital of the University, Dammam, Kingdom of Saudi Arabia. Tel. +966 (13) 8966666. Fax. +966 (13) 8966877. E-mail: a.s.alsaied@gmail.com

Disclosure. Authors have no conflict of interests, and the work was not supported or funded by any drug company.

Nodular fasciitis is a rare, benign reactive, or inflammatory self-limited condition of mesenchymal fibroblasts arising from the superficial or deep fascia or bony periosteum and presents as a rapidly growing mass.[1,2] Due to this pattern of growth, it can simulate a malignant lesion.[1,2] Nodular fasciitis was first described in 1955, and it was called subcutaneous pseudosarcomatous fibromatosis.[3,4] In addition, this lesion has been named proliferative fasciitis, subcutaneous fibromatosis, pseudosarcomatous fasciitis, and infiltrative fasciitis, as well as, being misclassified as several forms of sarcoma.[1,3,4] Nodular fasciitis can occur at any site of the body and it is equally frequent between males and females, the most common site of involvement is the upper extremity (48%), especially the volar aspect of the forearm, followed by the trunk (20%), head and neck (15-20%), and lower extremities (15%).[5,6] It accounts for 0.025% of all pathologic diagnoses, with less than 4% of those cases occurring in children aged 0-9 years of age,[3] and usually occurs in young and middle-aged adults.[5] This lesion frequently presents as a small mass with an average diameter of less than 3 cm, although lesions up to 10 cm have been reported in the literature.[7] Our objective in presenting this particular case is to report a 2-year-old boy, with a rare location of this lesion, to draw the attention of ENT surgeons to this rare condition.

Case Report. This 2-year-old boy presented to our clinic for evaluation of a facial swelling in the right maxillary area. The parents noticed a slight fullness at the right nasal sidewall, which progressed rapidly during a period of 2 months to reach a significant size. They denied any history of preceding trauma and there was no epistaxis, runny nose, fever, or constitutional symptoms. The medical and surgical history was unremarkable. Upon physical examination, there was a 4x4 cm hard mass, non-tender, at the right maxillary area and extending to the right nasal sidewall with an eggshell crackling on palpation. The right eye was intact; however, he could not open his right eye because of the effect of the mass. The right inferior turbinate was

pushed medially and blocking the right side; otherwise, normal looking mucosa (**Figure 1**). A CT scan and MRI revealed a right maxillary well-defined mass, measuring 4.5x4.1 cm (**Figure 2**). At this point, a malignant lesion was at the top of our differential diagnosis list due to the rapid growth rate. Subsequently, he underwent fine needle aspiration (FNA) for cytology examination, and the result was negative for malignant cells, with scattered multinucleated osteoclastic type giant cells along with aggregates of polymorphs in a background of red blood cells. At this point, brown tumor, or reparative granuloma was suspected. However, because of a normal parathyroid hormone level the brown tumor was excluded. As a result, he underwent a complete mass excision through a right sub-labial incision, and the mass was excised in 2 parts (**Figure 3**). Initially, an extensive approach, like Weber-Ferguson or mid-facial degloving approach was planned, however, because of a negative result for malignancy on FNA, we decided to start with a less extensive approach as a trial. We believe that the soft texture of the mass allowed the surgeon for complete removal via the sub-labial incision. The decision was made to leave the bowed face of the maxillary cavity for remodeling. Inferior antrostomy was carried out to ensure the ventilation of the maxillary sinus. The post-operative stay was uneventful. The result of the microscopic examination showed a moderately cellular lesion, comprising proliferation of spindly cells with round to oval nuclei, eosinophilic cytoplasm, and occasional typical mitosis embedded in variably collagenized stroma with focal myxomatous areas (**Figure 4**). Immunohistochemical stains were negative for Desmin, smooth muscle actin (SMA), muscle specific actin, CD-99, and S-100. However, it was positive for Vimentin, which was consistent with nodular fasciitis. Currently, he is on regular follow-up in our clinics for the last year, with no evidence of recurrence clinically or radiologically (**Figure 5**).

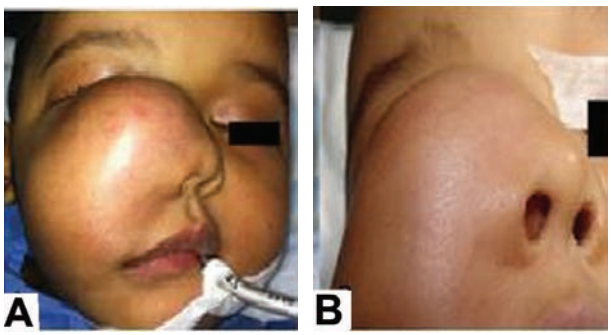

Figure 1 - An image of the patient with: right maxillary mass (**A**); and pushing of the right inferior turbinate (**B**).

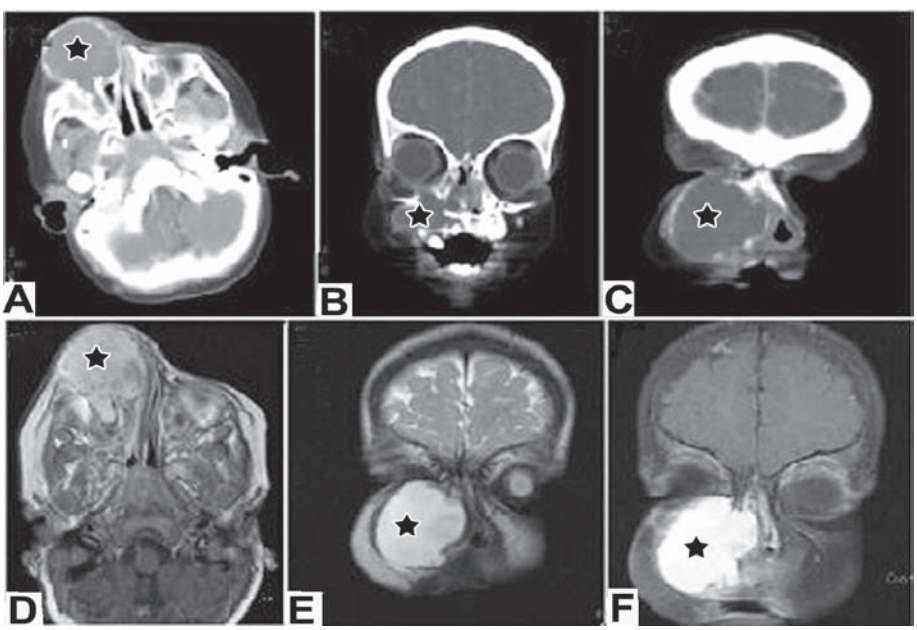

Figure 2 - Axial and coronal CT scan, post contrast administration showing: a non-enhanced right maxillary sinus mass (star)(**A**, **B**, & **C**); while in the axial and coronal MRI, the lesion appears to be low intensity in T1 (star) (**D**), high intensity in T2 (star) (**E**), and there was avid enhancement post gadolinium administration (star) (**F**).

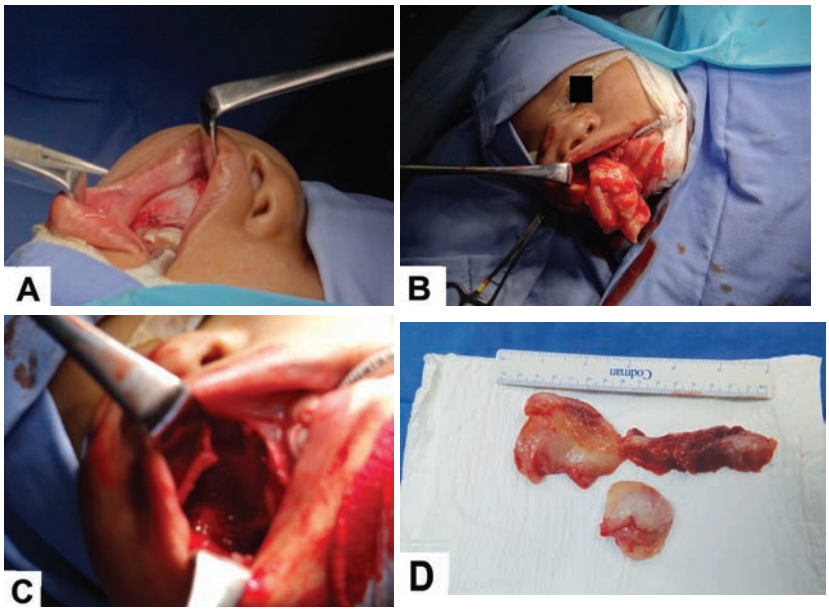

Figure 3 - Intra-operative pictures showing: **A**) right sub-labial incision; **B**) the mass being excised via a right sub-labial approach; **C**) the maxillary cavity after removal of the mass; **D**) a gross picture of the mass post-excision. The larger measuring 5.5x 5.0 x 3.5 cm, and the smaller measuring 4.2 x3.3 x 2.8 cm.

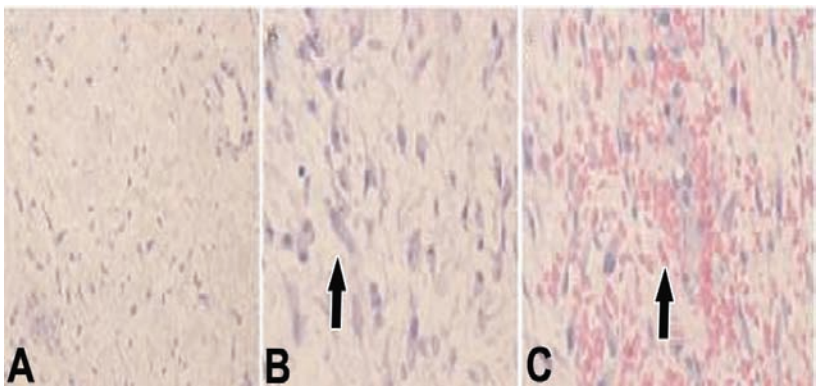

Figure 4 - Hematoxylin & Eosin stain showing: **A**) proliferation of fibroblast-like spindly cell with round to oval nuclei within loose and myxoid matrix, giving the tissue culture appearance; **B**) high magnified picture (arrow: fibroblast-like spindle cell); and **C**): extravasation of red blood cells (arrow).

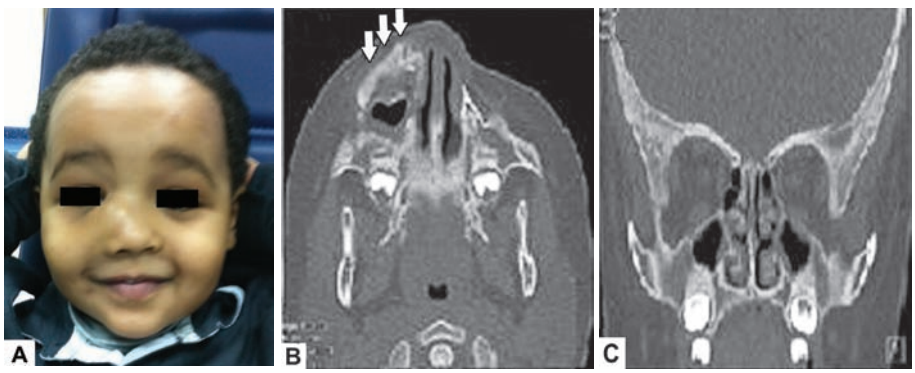

Figure 5 - An image showing: **A**) one year post-operative, appreciable remodeling process; **B**) axial CT scan showing a slightly bowed anterior wall of the right maxillary sinus (white arrows); **C**) coronal CT scan showing good ventilation of the sinus, and no signs of recurrence.

Discussion. The pathophysiology of nodular fasciitis is still not fully understood, though it is believed to be a fibroblastic, or myofibroblastic proliferation triggered by local trauma or inflammatory process.[2,3,5,6] However, if it is related to local trauma, it is difficult to explain why trauma is reported only in 10-15% of cases, and why the lesion is most common in the upper half of the body.[3] It typically presents as a painless mass; however, pain and/or tenderness are also possible upon presentation, which can be explained by the compression of the peripheral nerves by the lesion.[1] Macroscopically, nodular fasciitis is well circumscribed, but not encapsulated.[3,5] Microscopically, it is characterized by plump, immature-appearing fibroblasts, and a variable amount of mature collagen. Typical mitotic figures are common, which differentiate it from sarcoma as the latter has atypical mitosis.[4,5] Histologically, tissue culture appearance, extravasation of red blood cells, and the presence of more than one typical mitotic figure, especially in the myxoid sub-type, can be considered the most important diagnostic criteria for nodular fasciitis.[4] In 1961, nodular fasciitis was classified into 3 histological sub-types based on their predominant histological features: myxoid (type 1), which is the most common among them, and more common in children, cellular (type 2), the second in frequency, and fibrous (type 3), is the least common.[1,3-5] The myxoid sub-type is characterized by an abundant, nearly acellular central stroma, with more fibroblasts at the periphery. It shows significant inflammatory changes and a frequent red cell extravasation, which resemble a repair or granulation tissue. The cellular sub-type has a higher cellularity than the myxoid lesion. Whereas, fibrous type lesions are characterized by increased collagen production.[3] The immunohistochemical profile is substantial in diagnosing nodular fasciitis, this lesion shows positive reaction to Vimentin staining, which is a marker for mesenchymal cells. In addition, SMA could be positively stained, and it is generally more intense in the more cellular lesions. However, staining for S-100 and Desmin is usually negative.[3] Radiological studies of nodular fasciitis, weather CT scan or MRI, will reveal a relatively well-defined, soft-tissue mass of superficial location. Despite being histologically benign, the deep-seated lesions tend to be larger, with ill-defined margins, and may show an aggressive clinical behavior simulating a malignancy.[5] Typically these lesions are solid; however, cystic lesions have been reported.[5] Although various degrees of enhancement have been noted, moderate to strong enhancement has frequently been reported, and this could be because of the compact cellularity with a prominent capillary network.[5] Usually, nodular fasciitis on MRI will have isointense signal on T1-weighted images and hyperintense signal on T2-weighted images. However, the relationship between the signal intensities on T2-weighted images and histologic sub-types has been explained. Generally, the signal intensity of the lesions with myxoid or cellular sub-types is hyperintense on T2-sequence, whereas the signal intensity in fibrous sub-types lesions will be hypointense on all sequences. The coexistence of abundant collagen and acellularity leads to the reduction in signal intensity in the fibrous sub-type.[5] Because there are no specific diagnostic radiological findings for nodular fasciitis, there is a long list of differential diagnoses.[5] There is a histological transformation from type 1 to 2, then to 3. Thus, the histological appearance of nodular fasciitis may change in time from the active myxoid to the cellular, and lastly to the mature fibrous type, which will be reflected on the clinical course, radiological appearance, and the rate of growth. However, there is no clear explanation how these histological transformations occur in nodular fasciitis.[4]

The standard treatment is conservative complete surgical excision.[1-4,6] However, different modalities of treatment have been described in the literature. Graham et al[6] reported a forearm nodular fasciitis that was treated successfully with intra-lesional steroid (triamcinolone acetonide) injection, with fast and marvelous results.[6] However, a spontaneous regression after incisional biopsy of a nodular fasciitis in the zygoma has been reported also, even the signal intensity on T2-weighted imaging changed after the biopsy from a hyperintensity signal at the diagnosis time, to a hypointensity signal after 2 years follow up.[7] The recurrence rate after excision is 0.4-1%, but many authors believe that any recurrence indicates error in the initial diagnosis.[1-4] Bernstein and Lattes[8] recently stressed that recurrence of a lesion originally diagnosed as nodular fasciitis should lead to a careful re-evaluation of the pathological findings. In contrast, in the largest series of nodular fasciitis the authors reported a case of a palmer nodular fasciitis that was surgically excised, one month later the lesion recurred and was re-excised again, both lesions were proven histologically to be nodular fasciitis.[4] In addition, even a residual lesion has been reported to be stable with no evidence of progression during the follow up.[2]

We report a case of unusual nodular fasciitis, we believe it is unusual in several aspects. First, the age of presentation, since our patient is a 2-year-old. Secondly, the size of the lesion was more than the average size, which is 3 cm. Lastly, the location of the lesion, which is in the maxillary sinus. Our English literature review

(PubMed research), revealed a total of 3 published cases of nodular fasciitis in the para-nasal sinuses, only one of them was managed by sub-labial incision. Cotter et al reported the first case,[9] and they reported a complete excision of the left maxillary nodular fasciitis via the Weber-Ferguson approach. Hughes et al[10] reported the second case, and they described a sub-labial approach in the excision of left maxillary nodular fasciitis. Finally, Boyd et al[1] described a mid-facial degloving approach in resecting a nodular fasciitis arising from the left maxillary sinus.

In conclusion, although nodular fasciitis is a rare entity, this is the fourth case in the literature of nodular fasciitis located in the maxillary sinus. We believe that nodular fasciitis should be included in the differential diagnosis of a maxillary sinus mass. A sub-labial approach might be sufficient, with the best cosmetic results in managing this type of lesions.

References

1. Boyd ZT, Krad N, Nicklaus PJ, Lowe LH. Midfacial degloving approach to myxoid type nodular fasciitis of the maxilla in a 16-month-old female. ***Internet Journal of Pediatrics & Neonatology*** 2009; 10: 9.
2. Lenyoun EH, Wu JK, Ebert B, Lieberman B. Rapidly growing nodular fasciitis in the cheek of an infant: case report of a rare presentation. ***Eplasty*** 2008; 8: e30.
3. Haddad AJ, Avon SL, Clokie CM, Sandor GK. Nodular fasciitis in the oral cavity. ***J Can Dent Assoc*** 2001; 67: 664-667.
4. Shimizu S, Hashimoto H, Enjoji M. Nodular fasciitis: an analysis of 250 patients. ***Pathology*** 1984; 16: 161-166.
5. Kim ST, Kim HJ, Park SW, Baek CH, Byun HS, Kim YM. Nodular fasciitis in the head and neck: CT and MR imaging findings. ***AJNR Am J Neuroradiol*** 2005; 26: 2617-2623.
6. Graham BS, Barrett TL, Goltz RW. Nodular fasciitis: response to intralesional corticosteroids. ***J Am Acad Dermatol*** 1999; 40: 490-492.
7. Yanagisawa A, Okada H. Nodular fasciitis with degeneration and regression. ***J Craniofac Surg*** 2008; 19: 1167-1170.
8. Bernstein KE, Lattes R. Nodular (pseudosarcomatous) fasciitis, a nonrecurrent lesion: clinicopathologic study of 134 cases. ***Cancer*** 1982; 49: 1668-1678.
9. Cotter CJ, Finn S, Ryan P, Sleeman D. Nodular fasciitis of the maxilla in a child. ***J Oral Maxillofac Surg*** 2000; 58: 1447-1449.
10. Hughes C, Revington P, Berry J, Saunders M. Extracranial faciitis in childhood: a sheep in wolf's clothing. ***Br J Oral Maxillofac Surg*** 2002; 40: 341-343.