Case Report

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Nodular Fasciitis of Head and Neck in Childhood- A Chameleon Mimicking Sarcoma: Two Case Reports with Review of Literature

Neha Tyagi*, Akansha and Leelawathi Dawson

Dept. of Pathology, VMMC and Safdarjung Hospital, New Delhi, India

ABSTRACT

Nodular fasciitis is a benign, pseudosarcomatous proliferative lesion of the soft tissue which is frequently misinterpreted as sarcoma, both clinically as well as microscopically. Incidence of nodular fasciitis in head and neck region being 13% only. We report here two cases of nodular fasciitis both occuring in head and neck region.

Keywords: *Nodular Fasciitis, Benign, Sarcoma*

Introduction

Nodular fasciitis is a reactive, non-neoplastic lesion in which fibroblast like cells proliferate to form a fibrous mass and is thought to originate from muscular fascia(1). First reported in 1955 by Konwalla et al, who named it pseudo-sarcomatous fascitiis because it resembles sarcoma. The lesions most commonly occur in the upper extremities (43%),trunk(25%),lower extremities(22%) and head and neck region(13%).The incidence of nodular fasciitis peaks in 4th decade of life, with an approximately equal sex distribution(2).Nodular fasciitis is rarely diagnosed in childhood but appears in the head and neck region more commonly in children than in adults (3).The cause of nodular fasciitis is unknown but an association with the trauma may be present. Treatment is mostly by local surgical excision and recurrence is rare (4).

Case Report

CASE 1: A 16 year old female complaining of right cheek swelling since 6 months. It was a solitary, firm swelling about 1cm in size. Clinical diagnosis of infected sebaceous cyst was made. FNAC was done which yielded benign spindle cells in loosely adhesive cluster suggestive of a mesenchymal lesion. Soft tissue mass was excised and sent for histopathology. H&E sections showed spindle cells in bundles and fascicles along with lymphocytic infiltrate and extravasated RBCs (Fig 1a). Immunohistochemistry showed smooth muscle actin (SMA) (Fig 1(b)) and vimentin positivity . Therefore, a final diagnosis of Nodular fasciitis right cheek was given.

CASE 2: A 16 year old male presented to surgery OPD with an intraoral mass of 2X1cm which was soft, non tender. Clinical diagnosis of mucus retention cyst was made and excised mass was sent for histopathological examination. A globular, well encapsulated grey white soft tissue piece measuring 1.5X1X0.6cm was received which on cut surface was grey white homogenous. No cyst was identified grossly. H&E sections showed well circumscribed spindle cell lesion arranged in short fascicles forming storiform pattern. Few myxoid areas along with extravasated RBCs were also seen. Immunohistochemistry was positive for SMA and Vimentin. A diagnosis of Nodular Fascitis was made.

Discussion

Benign fibrous tumours represent a group of clinical entities that are often difficult to diagnose, nodular fasciitis being one such lesion (5). A variety of names have been given to these lesions including inflammatory pseudotumour, pseudosarcomatous fibro-myxoid tumour, pseudosarcomatous fasciitis, pseudosarcomatous fibromatosis, infiltrative fasciitis, postoperative spindle cell tumour and nodular fasciitis (6).

The most common localisations are the upper extremity (49%), trunk(18%), head and neck(13%) and lower extremity (17%) (7).Nodular fasciitis of the head and neck is rare in adults but more common in children. In fact, the head and neck region is by far the most common single site in children (8).Males and females are equally affected. It usually presents as a rapidlly growing soft tissue mass ranging from 0.4 to 10.5cm in diameter but usually not exceeding 3cm (9).

Although the cause of nodular fasciitis is unknown, it is speculated to be a reactive myofibroblastic proliferation initiated by a local trauma. The histological resemblance to organising granulation tissue support the theory of its origin. The term “fasciitis” implies that it is an inflammatory lesion originating in the fascial connective tissue surrounding blood vessels,nerves and muscles (10).

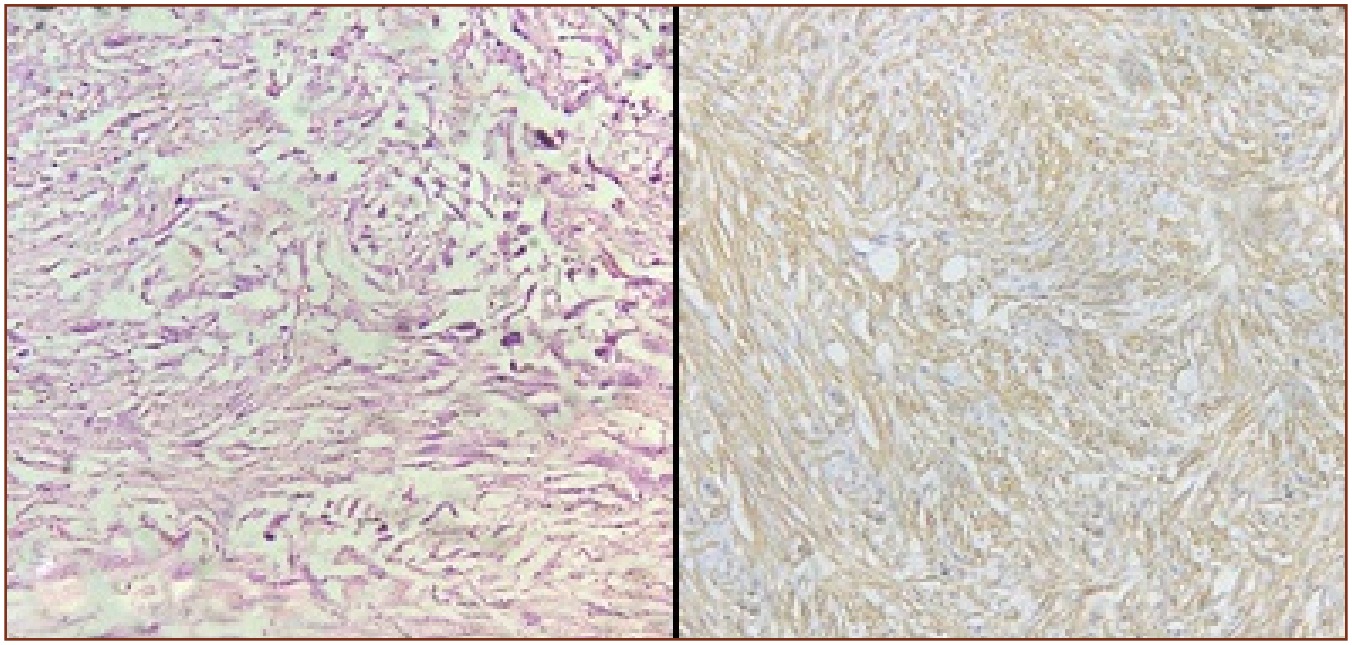

Fig. 1(a): H&E section (40X) showing a haphazard arrangement of spindle cells. Fig 1(b) Immunohistochemical staining (10X) showing Smooth Muscle Actin (SMA) positivity.

The lesion consist of nodular, encapsulated mass. The cut surface may show firm and grey white or soft and gelatinous areas. The subtypes are: Fascial, subcutaneous, intramuscular and intradermal type(4).

Histologically differential diagnosis may include proliferative fasciitis but in contrast with nodular fasciitis, it tends to be poorly circumscribed and often contains ganglion like cells.Other differential diagnosis include myxoma, fibrous histiocytoma and fibromatosis and can be differentiated by their growth pattern and cytologic features. Fibromatosis can furthur be distinguished by positive expression for beta catenin which is not expressed in nodular fasciitis (10).

A few malignant neoplasms can share histologic features with nodular fasciitis. Fibrosarcoma can also have active mitotic rate, but would be characterised by atypical mitosis and significant nuclear pleomorphism. Other spindle cell malignancies which might be included in the differentials include monophasic synovial sarcoma, leiomyosarcoma and malignant peripheral nerve sheath tumour(10). Therefore immunohistochemistry can be a useful tool to aid in the diagnosis.

The rapid clinical onset, presence of stromal chronic inflammation and lack of cytologic atypia are all critical features that can help identify the lesion as benign. On immunohistochemistry ,nodular fasciitis demonstrates vimentin, focal smooth muscle and smooth muscle actin, but not desmin, CD34 and S-100.The following features rule out malignant tumou (i) Absence of atypia, (ii) Absence of atypical mitotic figures, (iii) Small size, (iv) Short history and (v) Superficial location in adults (4).

Nodular fasciitis rarely recurs after conservative surgical excision is performed. Intralesional steroids have been tried with varied success. Recurrence after resection should raise the question of an alternative diagnosis (11).

Conclusion

The importance of recognising the true nature of fasciitis relates to the potential for overtreatment on the basis of microscopic features that are suggestive of malignancy. Both of these cases demonstrates that although infrequent in children, nodular fasciitis should be considered in the differential diagnosis of facial tumours for an accurate diagnosis and timely treatment.

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***Corresponding author:**
Dr Neha Tyagi, E 60 anand niketan new delhi 110021
Email: drnehatayagi@gmail.com

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