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An Unusual Presentation of Nodular Fasciitis: A Rare Case

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Abstract

Background: Nodular fasciitis is a benign soft tissue lesion caused by reactive proliferative fibroblast mainly according to literature triggered by trauma. Morphologically it mimics sarcoma hence also called pseudosarcoma. It is commonly seen in upper and lower extremities, as well as the trunk but rarely in the head and neck region. We are reporting a rare presentation of nodular fasciitis seen over right maxillary region.

Case Report: A 27-year-old chronic smoker male presented with 3-month history of gradual increasing in size of painless right maxillary mass with history of trauma to the face more than 6 months prior to presentation with no other significant history. On examination there was a swelling seen over right maxillary region measuring 3 cm x3cm, which was mobile with firm to hard in consistency, non-tender with no overlying skin changes. Computed Tomography of the paranasal sinuses showed a well-defined round heterogeneously enhancing subcutaneous soft tissue lesion seen at the right anterior maxillary region measuring 1.8 cm X 2.0 cm with no bony invasion, or calcification seen. Fine Needle Aspiration and Cytology revealed spindle cell lesion. Patient underwent tumor excision via sublabial approach and hhistologically reported as nodular fasciitis with positive for SMA (smooth-muscle specific actin) stain. Patient recovered fully postoperatively and no recurrence upon subsequent follow up.

Conclusion: In conclusion, although Nodular Fasciitis is uncommonly seen over head and neck region, it should always be considered as one of the differential diagnosis especially for any painless subcutaneous mass. A thoughtful consideration and careful work up should be done to obtain a proper diagnosis and treatment of this favorable lesion as the prognosis is good with low recurrence rate.

Keywords: Nodular Fasciitis; Fibroblast; Trauma; Pseudosarcoma

Background

Nodular fasciitis is a benign soft tissue lesion caused by reactive proliferative fibroblast mainly according to literature triggered by trauma.

Case Report

A 27-year-old male with no known underlying illnesses who is a chronic active smoker presented with swelling of right maxillary region which was gradually increasing in size for three months. Patient denied any pain, insect bite, nasal and dental symptoms, blurring of vision or weight loss. However, he had a positive trauma history over the face more than 6 months prior to presentation; in which the swelling has yet to appear. There was no family history of malignancy. On examination, he was comfortable and not in any distress, his vital signs were stable. There was a prominent swelling seen over right maxillary region measuring 3 cm x 3 cm, which was mobile with firm to hard consistency, non-tender with no overlying skin changes (Figure 1). Facial nerve and other cranial nerve were intact. No other swelling seen over facial or cervical region. No abnormalities seen via rigid nasoenodoscopy. The blood investigations were unremarkable.



Figure 1: Right maxillary swelling.

Computed Tomography (CT) of the paranasal sinus showed a well-defined round heterogeneously enhancing subcutaneous soft tissue lesion seen at the right anterior maxillary region measuring 1.8 cm X 2.0 cm with no fat or calcification component (Figure 2). The lesion was in continuity with the superficial facial fascia with minimal scalloping of the adjacent anterior wall of right maxillary sinus. No erosion or extension into the maxillary sinus and no involvement of the adjacent skin anteriorly or stranding of the surrounding fat. Fine needle aspiration and cytology was done cytomorphological features favor spindle cell lesion. Patient underwent tumor excision via sublabial approach under general anesthesia. Intraoperatively, a 2 cm by 2 cm mass seen at subcutaneous of right maxillary region with no bony erosion seen. Histopathological examination showed tissue consists of spindle shaped cells arranged in "C" and "S" shaped short fascicles and storiform pattern, set in myxoid stroma. The spindle cells have plump ovoid to elongated vesicular to hyper chromatic nuclei with dispersed chromatin, visible nucleoli, bipolar pale eosinophilic cytoplasmic processes with ill-defined cellular border (Figure 3). The mitotic count is 1 per 2 mm square. No aberrant mitosis or necrosis seen. Microcystic stromal change is observed in areas. Red blood cells extravasation were seen as well with scattered lymphocyte infiltrates seen throughout the lesion. There are also ectatic to branching thin-walled vascular channels within the lesion. The lesional cells are positive for SMA and negative towards CD34 and EMA. Post-operatively patient was given one week dose of antibiotic and 3 doses of Dexamethasone and showed complete recovery (Figure 4).



Figure 2: Computed tomography of paranasal sinus (Axial view).

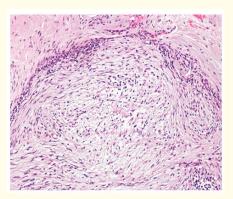


Figure 3: Proliferation of spindle-shaped cells in "C" and "S" shape short fascicles and storiform pattern. Microcystic stromal changes accompanied by areas of haemorrhage.

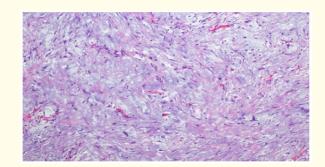


Figure 4: The spindle cells have plump ovoid, elongated vesicular to hyperchromatic nuclei with dispersed chromatin, bipolar pale eosinophilic cytoplasmic processes and ill-defined cellular border. Extravasation of RBCs is seen in the background



Figure 4: One month post operation.

Discussion

Nodular fasciitis (NF) or originally described as subcutaneous pseudosarcomatous fibromatosis is a benign tumor-like fibroblastic proliferation lesion. It is commonly seen in upper extremities (39% - 54%), the trunk (15% - 20%); the lower extremities (16% - 18%), and head and neck (7% - 20%). NF usually affects those in the 20s - 40s regardless of gender [1]. The aetiology of NF is not entirely known but it has been said that antecedent trauma may be a triggering factor; however, it is not being widely accepted [2]. Typically, it presents as firm subcutaneous or submucosal mass that grows insidiously from the underlying muscular fascia. Commonly it is seen < 4 cm in size however the size may vary from 0.5 cm - 10 cm [3]. According to anatomic location NF can be divided into three; subcutaneous, intramuscular, and intermuscular with subcutaneous being the commonest by 3 - 10 fold. On the basis of type extracellular matrix, NF can be class into myxoid, cellular and fibrous type. NF is often mistaken with connective or soft tissue tumor such as sarcoma, especially when they both may present with similar history of rapid growth. Radiologically features of NF are rather nonspecific. Commonly a relatively well-defined, moderate to strong enhancement superficial soft-tissue mass can be seen with either Computed Tomography or Magnetic Resonance Imaging. Hence histopathological and immunohistochemistry comes in handy in giving an accurate diagnosis. Histologically NF may mimic sarcoma but are differentiated by immature-appearing fibroblasts. Fusiform proliferated cells arranged in short S-shaped fascicles within a myxoid matrix, with many small vessels and some erythrocytes. Cellular atypia is uncommon despite high mitotic index.

Further staining is the needed to rule out other malignancies. In NF, cells are usually positive for SMA staining and a negative expression towards CD34 and EMA may rule out sarcoma and meningioma respectively⁴. As seen in our case, patient's immunohistology findings were exactly as described in literature. Patient with NF may have spontaneous regression and which then will continue with regular follow up or observation. However, in bigger mass or lesion that has other associated symptoms (i.e. neurological deficit or movement limitation etc.), an intra-lesional steroid injection or surgical excision may be done as the prognosis is good with very rare occasion of recurrence [4].

Conclusion

In conclusion, although nodular fasciitis is uncommonly seen over head and neck region, it should be considered as one of the differential diagnosis especially for any painless subcutaneous masses with a history of predisposing trauma. A thoughtful consideration and careful work up should be done to obtain a proper diagnosis and treatment of this favorable lesion as the prognosis is good with low recurrence rate.

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77