Case Report

Oral Solitary Neurofibroma with Ossification

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Abstract

Solitary neurofibroma is a benign neoplasm of neurogenic origin i.e. Schwann cell and perineural fibroblasts. We report a case of solitary neurofibroma in the oral cavity with the unusual occurrence of osseous metaplasia within the tumour. It is important for the pathologist to recognize these secondary features of metaplastic bone formation in the soft tissue tumours to avoid erroneous diagnosis.

Key Words: Neurofibroma, Neural tumour, Ossification, Schwann cell

Introduction

Neurofibroma is a benign neoplasm of peripheral nerve believed to be originating from mixture of cell types that includes schwann cell and perineural fibroblasts[1]. Solitary neurofibroma within the oral cavity is relatively a rare occurrence, despite being one of the most common neurogenic tumours.

Clinically it may present either as solitary lesion or be a component of syndrome called neurofibromatosis that generally presents with multiple lesions on the skin[1]. Its characteristic histopathological features include proliferating spindle cells which exhibits wavy nuclei in a delicate fibrous stroma[1]. Diffuse distribution of mast cells may be seen which often helps in accurate diagnosis of this neoplasm[1].

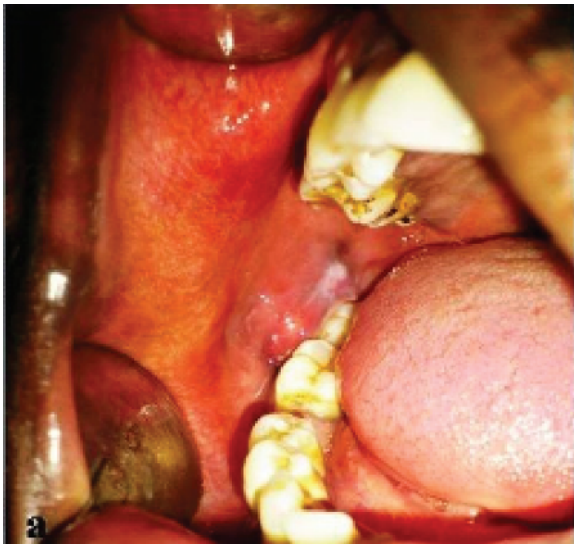
Here we report a case of an intraoral solitary neurofibroma which presented with extensive metaplastic bone formation within the tumour. Till to date, totally three cases of neurofibroma with ossification have been reported in the literature, of which two are solitary neurofibroma[2,3] and the other one is diffuse neurofibroma[4]. To the best of our knowledge, this is the first case of oral solitary neurofibroma to be reported in the literature with this unusual histopathological feature.

Case description

A 40 year old male patient presented with the chief complaint of painless, gradually progressive swelling in the right posterior cheek region of two month duration. Clinical examination revealed a well-defined solitary nodule obliterating the mandibular right buccal vestibule in relation to 47 and 48 region. It was of size approximately 3 x 2.5 cm, with colour similar to adjacent mucosa except for focal whitish appearance in the posterior aspect of the swelling caused by impingement of the opposing maxillary teeth. No surface changes evident [Fig 1A]. Palpation around the periphery of the solid mass revealed a well defined smooth border and the lesion was found to be freely mobile (sub mucosal). It was hard in consistency and was not tender on palpation.

Mandibular occlusal and right lateral oblique radiograph revealed no evidence of any distinctive radio-opacity in the region of swelling. Based on the clinical and radiographic findings a provisional diagnosis of solitary soft tissue tumour was made with different possibilities such as fibroma, schwannoma, solitary fibrous tumour etc.

Surgical excision of the lesion was planned under local anaesthesia. Superficial mucosal incision was made using electrocautery. Subsequent reflection of the mucosa revealed a firm, whitish, roughly ovoid solitary lesion without any attachment to the underlying periosteum or the jaw bone [Fig 1B].



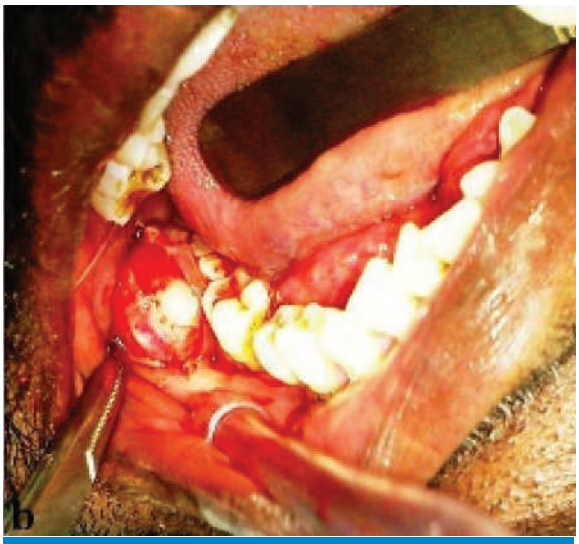


Fig 1 : Sub mucosal nodule in the mandibular buccal vestibule in relation to 47 & 48 (a) and surgical exposure of the lesion after mucosal reflection (b)

The lesion was stabilised with suture and shelled out from the surrounding tissue. Grossly the specimen appeared to be solitary solid mass, white in colour, with smooth surface and measured about 2 x1.5 x 1 cm [Fig 2A] Cut surface was found to be white to yellow-ish-white in colour and firm on palpation [Fig 2B].

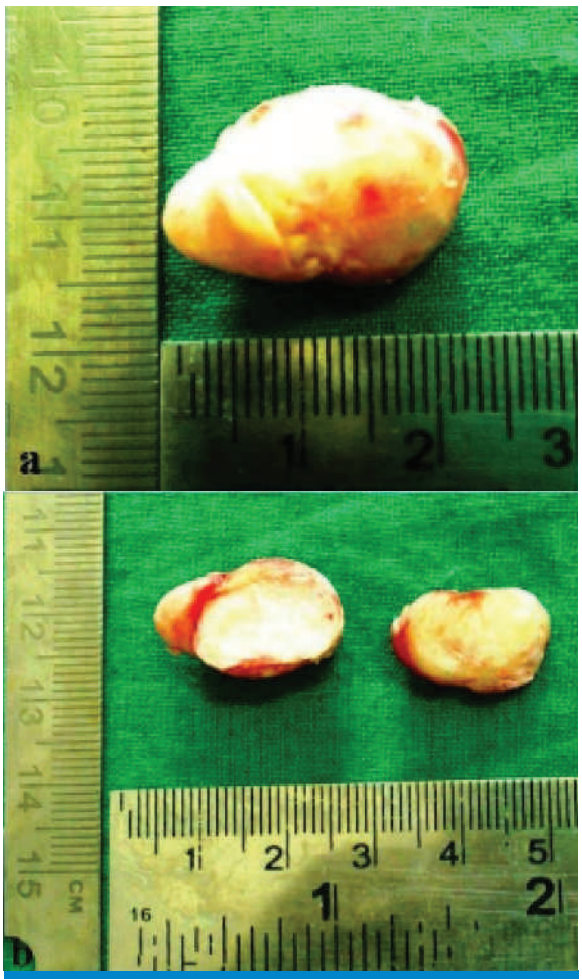


Fig 2 : Nodule as a whole after removal from the surgical site (a) and the cut surface (b)

Histopathological examination of the haematoxylin and eosin (H&E) stained sections basically showed fibro-cellular connective tissue stroma composed of areas of more dense fibres and loosely arranged delicate collagen fibres, with associated spindle cells exhibiting wavy nuclei [Fig 3].

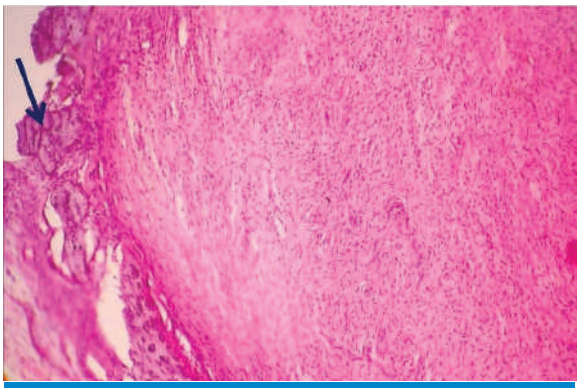


Fig 3 : Spindle cells exhibiting wavy nuclei associated with delicate collagen fibrils with associated ossified areas (arrow) (H&E 10 X)

Distinctive bone formation or ossification with varying degree of maturation, peripheral osteoblastic rimming and entrapped osteocytes was seen diffusely in the fibro-cellular stroma [Fig 4a]. Toluidine blue staining revealed scattered distribution of mast cells in the tumor stroma [Fig 4b].

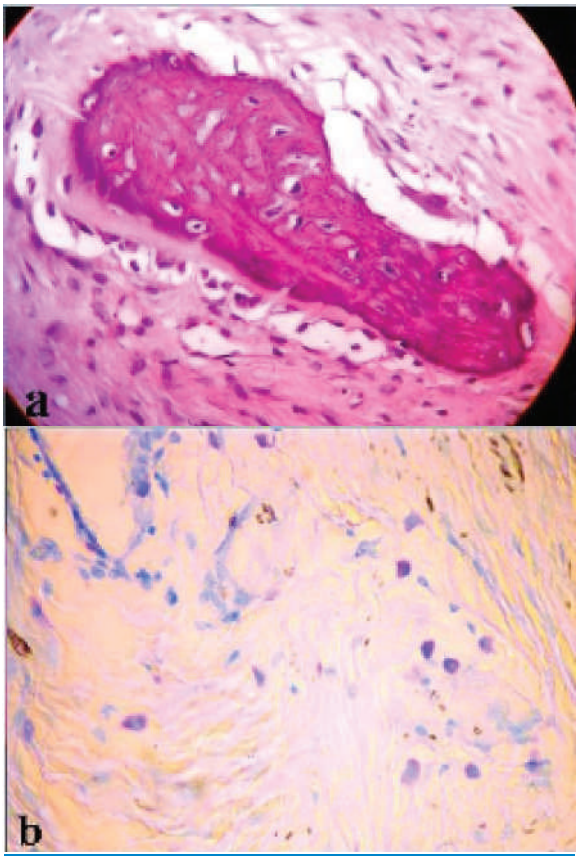


Fig 4 : Osseous metaplasia with osteoid, osteoblastic rimming and entrapped osteocytes H&E 40X (a) and scattered mast cells stained purple (metachromatically) within the tumour stroma (Toluidine blue 40 X) (b)

Immunohistochemical investigation utilizing antibody for S-100 protein antigen (Dako-IS-504, Denmark,) showed diffuse positive reaction [Fig 5].

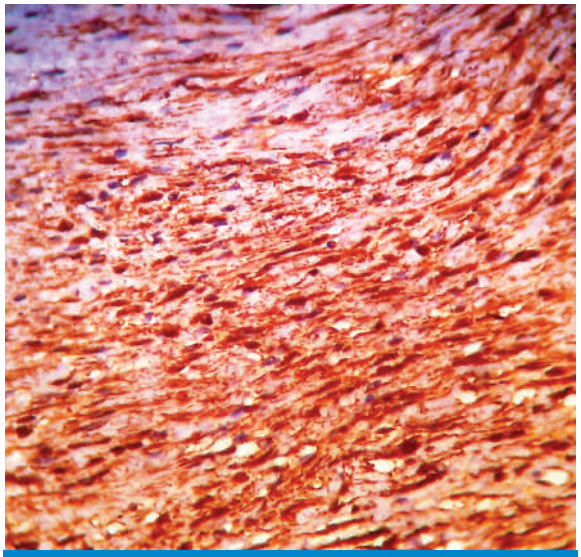


Fig 5 : Immunohistochemical positivity for S-100 protein antigen

Based on the clinical, histopathological and immunohistochemical features, a final diagnosis of solitary neurofibroma with metaplastic bone formation was made. One and half year follow up patient showed no evidence of recurrence.

Discussion

Among the neurogenic neoplasms, neoplasm of Schwann cell origin is the most frequent occurrence and mainly includes neurilemmoma, solitary neurofibroma, neurofibroma in neurofibromatosis syndrome and malignant peripheral nerve sheath tumour[1].

The present case is diagnosed as solitary neurofibroma based on the clinical, characteristic histopathological features and immunohistochemistry, that were described in case report section. Presence of diffuse metaplastic bone formation or ossification represents an additional uncommon histopathological finding in the present case. It has been theorized that, Schwann cells are pleuripotential cells and have the ability of metaplastic bone and cartilage formation[2]. This nature has been already evident from the occurrence of metaplastic bone, cartilage and muscle in various forms of malignant peripheral nerve sheath tumor[5].

Being a neoplasm of Schwann cell origin, the above mentioned fact, explains the occurrence of osseous metaplasia in neurofibroma. But dearth of available literature 2-4 reporting neurofibroma with metaplastic bone formation explains its rarity, despite it being one of the most common neurogenic neoplasms.

The clinical implication of this secondary phenomenon is that, occasionally this form of metaplastic bone formation may obscure the features of primary lesion and can lead to an erroneous diagnosis of primary osteogenic tumors.

Absolute bland cytological features of the neoplastic cells, lack of atypical mitotic figures and well defined borders in the histopathology, ruled out the malignant transformation in the present case.

Conclusion

Till date there has been no report of solitary neurofibroma showing metaplastic bone formation from the oral and maxillofacial region which justifies its documentation for future reference. This case highlights and provides practical evidence for osteoblastic differentiation ability of perineural cells i.e Schwann cells and perineural fibroblasts. It also emphasizes the importance of distinguishing the secondary phenomenon of osseous metaplasia in various soft tissue tumors from the primary osseous pathologies in the soft tissue by the pathologist. The prognostic significance if any associated with the presence of osseous metaplasia needs further documentation and follow-up of such cases.

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