Myofibroma of Mandible: A Diagnostic Dilemma
Purwa Patil, Bhavana Bharambe, Aditi Beke

Abstract
Intraosseous myofibroma is a rare tumour characterised by localised or generalised proliferation of fibroblastic tissue. Bony lesions are common in mandible and maxilla. Histopathology revealed proliferation of streaming and whorled fascicles of spindle cells around slit-like vascular spaces in collagenous stroma. The spindle cells were immunopositivity for vimentin and smooth muscle actin, but were negative for desmin and S-100 protein. Myofibroma appears alarming clinically due to its fast growth which may mimic a malignancy. However, it is completely benign and is usually treated by complete surgical excision with excellent prognosis. This tumor was positive for vimentin and SMA. Our patient has shown no signs of recurrence during 2 years of follow-up.

Keywords: Myofibroma; Mandible; Benign Tumor; Intraosseous; Biphasic Tumor; Myofibroblast.

Case Report

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Introduction
Myofibroma, as the name implies, is a benign tumour of the myofibroblasts commonly found in the head and neck region. The presentation is usually of an intradermal and soft tissue mass with equal involvement of male and females. Intraosseous lesions are rare relatively common in younger age group than adults. The swellings are frequently rubbery or firm, scar-like consistency with a size averaging from 0.5 to 1.5 cm. The symptoms are variable depending on the location of the tumour like respiratory distress, vomiting, or diarrhoea, sometimes proving fatal. Radiographically, it appears as a well-defined lesion with unilocular radiolucency. Histologically, well defined multinodular tumour arising in the deep dermis or subcutis with stag horn pattern of blood vessels interspersed within the tumour. We report a case of myofibroma featuring its clinical, radiological, histopathological and immunohistochemical features and the dilemma in its diagnosis histologically.

Case summary
An 18 year old female presented with swelling over left angle of mandible which had gradually increased over period of one year. Her medical history was non-contributory. On local examination, a fixed, non-tender swelling of 4x4 cm dimension was palpated over left side of mandible. The cone beam computed tomography of mandible revealed a well-defined, expansile, multiloculated, lytic lesion arising from left mandible involving body, ramus and coronoid process. Lesion was multiloculated causing scalloping of adjacent bony margins and displacing the left superior alveolar arch medially (Figure 1a). Lesion showed areas of demineralisation with cortical breach in continuity along the lateral and medial aspect. Walls of paranasal sinuses and nasal septum were intact. Radiologically dentigerous cyst or ameloblastoma was suspected. Hemi-mandibulectomy with removal of body, ramus and coronoid process was done with 1 cm wide margin. On gross examination, a nodular, greyish-white, solid mass of 4x4 cm was seen involving the body and angle of mandible. Molar tooth was also present (Figure 1b). Histologically, a well circumscribed tumour with nodular architecture was noted (Figure 1c). It comprised of short fascicles and whorls of plump, spindle shaped cells with tapering vesicular, nuclei and conspicuous nucleoli (Figure 1d). Hyalinised stroma showed pseudochondroid appearance at places with haemangiopericytomatos pattern of vessels. Stromal collagenisation with calcification was present. However, mitosis, necrosis or nuclear pleomorphism was not evident. Immunoreactivity for vimentin and smooth muscle actin was observed (Figure 1e & f).

Discussion
The myofibroma is a benign mesenchymal tumour frequently observed in the skin and subcutaneous tissue of the head-neck region. It was first described by Stout et al., in 1957. It can be infantile or adult depending upon the age of presentation. It is described mainly in infants with congenital forms and, in early childhood, with acquired...
forms. Less often, it can be observed in adolescents and adults as in our case. However, aetiology remains unknown. Soft tissue lesions are much more common than intraosseous ones like the mandibular involvement in the case discussed here. Rarely, systemic involvement of gastrointestinal tract, lung, heart and pancreas has been described. Clinical presentation of the patient is either with a mass or pain. On radiological examination, mandibular tumours have been described as well-defined either unilocular or multilocular radiolucencies with marginal sclerosis.

Figure 1: The Multiloculated lesion causing scalloping of bony margins with cortical breach in continuity along the lateral and medial aspect (a). The macroscopic examination revealed nodular, greyish-white, solid mass involving the body and angle of mandible (b). The hematoxylin and eosin stained section low power view showed streaming and whorled fascicles of spindle cells around hemangiopericytomatosus pattern of vascular spaces in collagenous stroma with pseudochondroid appearance (inset) (c & d). The immunohistochemical staining of myofibroblasts showed strong cytoplasmic positivity for Vimentin (e) and for smooth muscle actin (f).

<table>
<thead>
<tr>
<th>Tumour</th>
<th>Histological feature</th>
<th>Immunohistochemistry</th>
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<tr>
<td>Leiomyoma</td>
<td>No hemangiopericytomatosus areas</td>
<td>SMA, Desmin positive</td>
</tr>
<tr>
<td>Solitary fibrous tumor</td>
<td>Hemangiopericytomatosus areas seen, hyper and hypocellular areas with collagenisation</td>
<td>CD34, CD99</td>
</tr>
<tr>
<td>Nodular fascitis</td>
<td>No hemangiopericytomatosus areas</td>
<td>Desmin positive</td>
</tr>
<tr>
<td>Neurofibroma</td>
<td></td>
<td>S 100 positive</td>
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<tr>
<td>Fibrous histiocytoma</td>
<td>Polymorphous cells arranged in storiform pattern</td>
<td>SMA</td>
</tr>
<tr>
<td>Infantile fibromatosis</td>
<td>Monomorphic proliferation of spindle cells, no hemangiopericytomatosus areas</td>
<td>Vimentin positive</td>
</tr>
<tr>
<td>Myofibrosarcoma</td>
<td>Histologically features may overlap</td>
<td>SMA, Desmin positive</td>
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Table 1: Differential diagnosis of myofibroma

As the mandible is an uncommon location of a myofibroma, differential diagnosis must be established with other benign and malignant neoplasms, high or low grade. The histology of the myofibroma shows a biphasic growth pattern: elongated spindle cells with
eosinophilic cytoplasm, in the borders, polygonal cells arranged in a palisading pattern, with hyperchromatic nuclei, in the central portions. Depending on the predominance of cellular type, there can be a variety of lesions that come into its spectrum of differential diagnosis1,8, namely, nodular fascitis, fibrous histiocytoma, neurofibroma, leiomyoma, and infantile fibromatosis causing confusion in reaching the final diagnosis in many instances3,5 (Table 1). The diagnosis of myofibroma, usually made after excision, is obtained by means of immunohistochemistry, in which there is positivity for vimentin and α smooth muscle antibodies and negativity for keratin, S-100, EMA (Epithelial Membrane Antigen) and desmin antibodies.4 The treatment is surgical; the prognosis is generally good with low rates of recurrence after excision.4,8

Conclusion
Myofibroma is a benign fibrous tissue tumor which requires an integrated clinical, histopathological and immunohistochemical support for diagnosis. Over diagnosis and aggressive treatment of the lesion can be prevented by proper familial history, searching for multiple sites for the tumor and the use of ancillary techniques.

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