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原文作者姓名：	Purwa Patil, Bhavana Bharambe, Aditi Beke
通訊作者學校：	JJ Hospital Campus, Byculla,
報告者姓名(組別)：	鄭翊宏 Intern E 組
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內文：

#### Introduction

##### Myofibroma

- ① a benign tumour of the myofibroblasts commonly found in the head and neck region.
- ② an intradermal and soft tissue mass equal involvement of male and females.
- ③ Intraosseous lesions are rare relatively common in younger age group than adults.
- ④ rubbery or firm, scar-like consistency with a size averaging from 0.5 to 1.5 cm.
- ⑤ The symptoms are variable depending on the location of the tumour like respiratory distress, vomiting, or diarrhoea, sometimes proving fatal.
- ⑥ Radiographically, it appears as a well-defined lesion with unilocular radiolucency.
- ⑦ Histologically, well defined multinodular tumour arising in the deep dermis or sub-cutis with stag horn pattern of blood vessels interspersed within the tumour.

#### Case summary

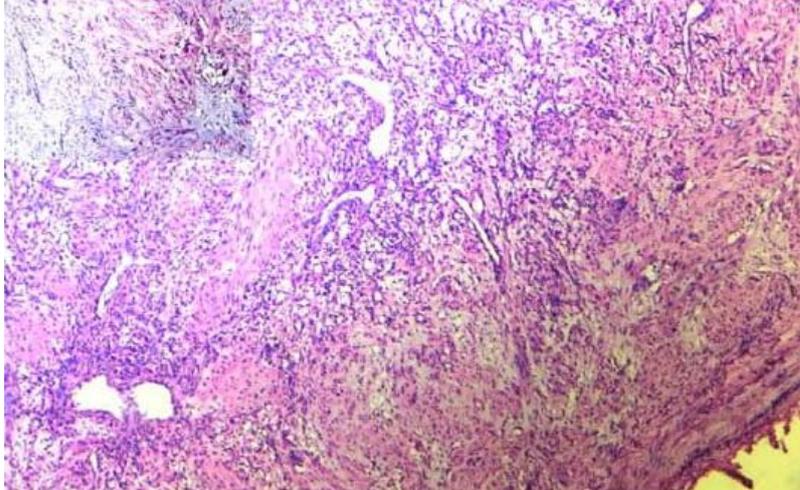
- ✓ An 18 year old female presented with swelling over left angle of mandible which had gradually increased over period of one year.
- ✓ Her medical history was non-contributory.
- ✓ On local examination, a fixed, non-tender swelling of 4x4 cm dimension was palpated over left side of mandible.
- ✓ The 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



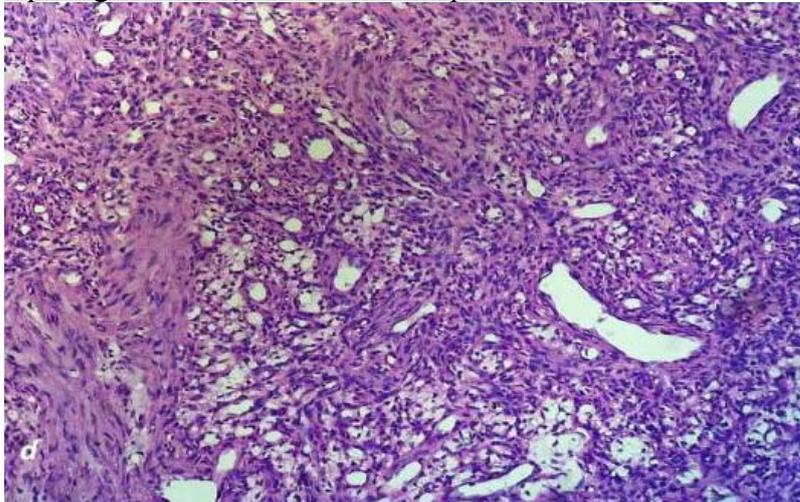
- ✓ Lesion showed areas of demineralisation with cortical breach in continuity along the lateral and medial aspect.
- ✓ Walls of paranasal sinuses and nasal septum were intact.
- ✓ Radiologically dentigerous cyst or ameloblastoma was suspected.
- ✓ Hemi-mandibulectomy with removal of body, ramus and 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



✓ Histologically, a well circumscribed tumour with nodular architecture was noted



✓ It comprised of short fascicles and whorls of plump, spindle shaped cells with tapering vesicular, nuclei and conspicuous nucleoli

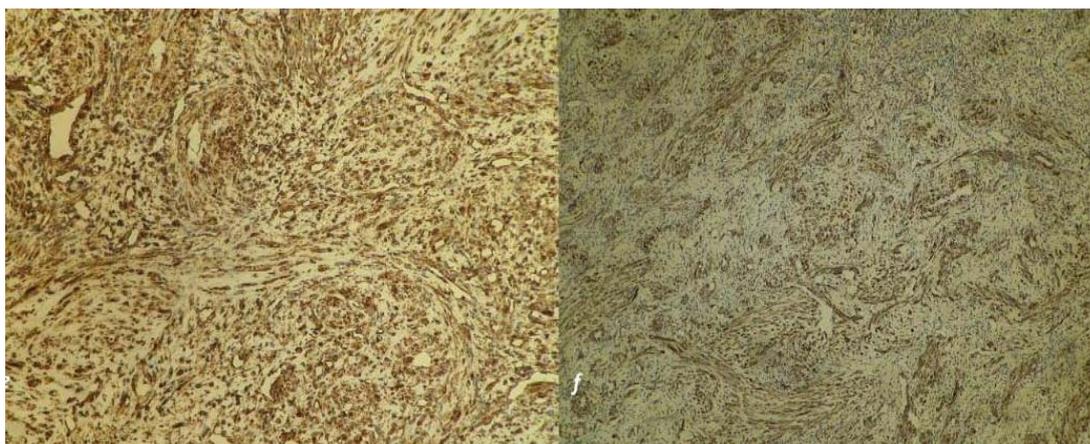


✓ Hyalinised stroma showed pseudochondroid appearance at places with haemangiopericytomatous pattern of vessels.

✓ 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

❶ The myofibroma is a benign mesenchymal tumour frequently observed in the skin and subcutaneous tissue of the head-neck region.

❷ It was first described by Stout et al., in 1957.

❸ 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.

❹ However, aetiology remains unknown.

❺ Soft tissue lesions are much more common than intraosseous ones like the mandibular involvement in the case discussed here.

❻ Rarely, systemic involvement of gastrointestinal tract, lung, heart and pancreas has been described.

❼ Clinical presentation of the patient is either with a mass or pain.

❽ On radiological examination, mandibular tumours have been described as well-defined either unilocular or multilocular radiolucencies with marginal sclerosis.

❾ 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.

✓ The diagnosis of myofibroma, usually made after excision, is obtained by means of immunohistochemistry, in which there is positivity for vimentin and  $\alpha$  actin smooth muscle antibodies and negativity for keratin, S-100, EMA (Epithelial Membrane Antigen) and desmin antibodies.

✓ The treatment is surgical; the prognosis is generally good with low rates of recurrence after excision.

 Table 1: Differential diagnosis of myofibroma

Tumour	Histological feature	Immunohistochemistry
Leiomyoma	No hemangiopericytomatous areas	SMA, Desmin positive
Solitary fibrous tumor	Hemangiopericytomatous areas seen, hyper and hypocellular areas with collagenisation	CD34, CD99
Nodular fasciitis	No hemangiopericytomatous areas	Desmin positive
Neurofibroma		S100 positive
Fibrous histiocytoma	Polymorphous cells arranged in storiform pattern	SMA

Infantile fibromatosis	Monomorphic proliferation of spindle cells, no hemangiopericytomatous areas	Vimentin positive
Myofibrosarcoma	Histologically features may overlap	SMA, Desmin positive

 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.

題號	題目
1	Immunoreactivity for myofibroma will show a positive result except ? (A) Vimentin (B) SMA (C) Desmin (D) S100
答案(CD)	出處：Myofibroma of Mandible: A Diagnostic Dilemma
題號	題目
2	A 16-year-old male patient reported with a chief complaint of swelling on the right side of the face for the past 2 months. On palpation the swelling was firm, nontender. CT scan revealed an osteolytic lesion about 2.5 cm × 2 cm in dimension, extending from the distal aspect of 47 to the ramus of the mandible. Destruction of buccal cortical plate was evident. Microscopic evaluation of the sections revealed interlacing fascicles of spindle-shaped cells arranged in a biphasic pattern set in a collagenous stroma. Vascular spaces mimicking the hemangiopericytoma pattern were also observed. Positive immunoreactivity was observed for vimentin and αSMA and negative immunoreactivity for S100, desmin, and CD68. What is the most likely tumor in this case ? (A) Myofibrosarcoma (B) Myofibroma (C) Leiomyoma (D) odontogenic keratocyst
答案(B)	出處：Myofibroma of Mandible: A Diagnostic Dilemma