CASE REPORT

Intraosseous myofibroma of mandible: A rarity of jaws: With clinical, radiological, histopathological and immunohistochemical features

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ABSTRACT

Myofibroma is an uncommon benign mesenchymal neoplasm composed of myofibroblasts, but it can be confused with more aggressive spindle cell tumors. Solitary myofibroma is common in soft tissues of head and neck, but rare in the jaw bones with only 38 cases of central myofibroma of mandible reported in English medical literature. When encountered in the jaws, lesions exhibit clinical and radiographic features suggestive of odontogenic cysts/tumors or other neoplastic conditions. We hereby present the 39th case of intraosseous myofibroma of the mandible which had been reported to our institution. A 16-year-old male reported with a chief complaint of swelling in the right side of face. Intraorally there was a firm, nontender swelling in the right buccal aspect of the mandible. Radiologically the lesion was osteolytic, destroying the buccal cortical plate. Histologically, characteristic biphasic pattern of myofibroma was noticed. Immunoreactivity was positive for vimentin and αSMA but negative for desmin, thus confirming our diagnosis. The patient was treated by local-wide surgical excision of the lesion. A 3-year follow-up revealed no signs of recurrence. Occurrence of myofibroma involving the jaw bones is common in the younger age groups and represents a unique diagnostic and therapeutic challenge. Differentiating this lesion from other benign and malignant neoplasms is crucial in deciding between a radical and a conservative treatment approach.

Key words: **Intraosseous, myofibroma, mandible, myofibroblasts**

INTRODUCTION

Myofibroma and Myofibromatosis represent an uncommon group of lesions that were described as congenital multicentric fibroblastic proliferation by Stout[1] who introduced the term "congenital generalized fibromatosis." The terms myofibroma (solitary) and myofibromatosis (multicentric) were adopted by WHO to describe the benign neoplasms of contractile myoid cells arranged around thin walled vessels.[2]

Recent reports on myofibromas demonstrate a predilection for the head and neck region, particularly oral and perioral structures in both solitary and multicentric patterns. Intraosseous cases especially in jaws are uncommon. Only a few sporadic cases of solitary myofibroma of mandible have been described. A literature survey revealed only 38 cases of myofibroma involving the mandible. The unique feature of central myofibroma of the jaws is its potential to involve the teeth or other odontogenic structures and exhibit alarming clinical or radiographic features suggestive of an odontogenic cyst/tumor or other nonodontogenic lesions.[3]

In this article we present the 39th case of solitary myofibroma of mandible in a 16-year-old male patient with detailed description of clinical, radiographic, histopathological, and immunohistochemical findings.

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CASE REPORT

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 [Figure 1]. The swelling was initially noticed

Figure 1: Clinical picture showing swelling in the right side angle of mandible

as a small lump which gradually increased to the present size. Medical and family histories were noncontributory. Examination revealed swelling in the right side of the face about 1.5 × 2 cm in size in the region of angle of mandible. There was no draining sinus and the skin over the swelling was clinically normal.

Intraorally the swelling was present in the buccal aspect of right mandibular second molar measuring about 1.5 cm × 1 cm in dimension. Mucosa over the swelling was clinically normal. Buccal cortical bone expansion was seen along with obliteration of buccal sulcus. On palpation the swelling was firm, nontender and extended superiorly to involve the anterior border of ramus of the mandible.

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. The clinical and radiologic findings were suggestive of odontogenic tumor [Figures 2a-c].

Subsequently, an incisional biopsy was performed under local anesthesia and the specimen was sent for histopathologic examination. Microscopic evaluation of the sections revealed interlacing fascicles of spindle-shaped cells arranged in a biphasic pattern set in a collagenous stroma. Spindle cells were seen with oval, round and tapering nuclei with pale eosinophilic cytoplasm. These cells were seen alternating with closely packed cells with small rounded nuclei and eosinophilic cytoplasm [Figure 3]. Vascular spaces mimicking the hemangiopericytoma pattern were also observed [Figure 4]. Cellular atypia was not noted. Based on these findings we arrived at a diagnosis of myofibroma.

Immunohistochemical staining was carried out for vimentin, S100, αSMA, CD68, and desmin. Positive immunoreactivity was observed for vimentin and αSMA [Figures 5 and 6] and negative immunoreactivity for S100, desmin, and CD68, thus confirming the myofibroblastic nature of the tumor. Further medical and radiographical examination confirmed the solitary nature of the lesion, thus excluding myofibromatosis. Hence

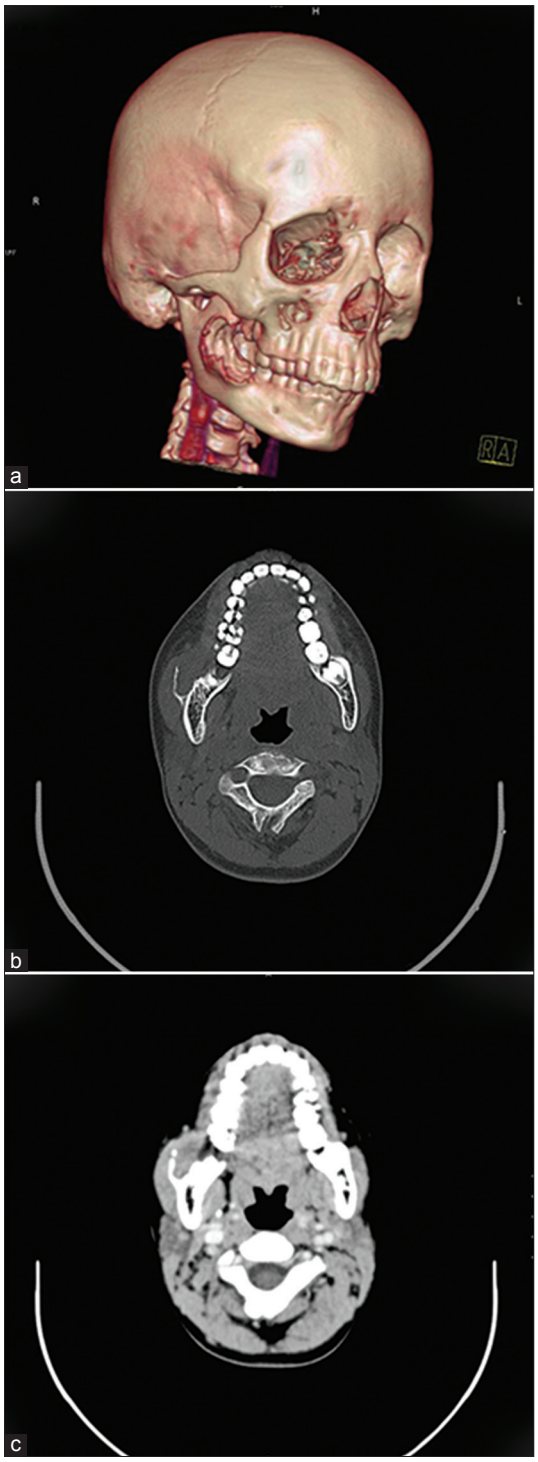

Figure 2(a-c): CT scan showing an osteolytic lesion about 2.5 × 2 cm in dimension extending from the distal aspect of 47 to the ramus of the mandible with destruction of buccal cortical plate

local-wide excision of the lesion was performed under general anesthesia. Histopathologic findings of the postsurgical specimen also confirmed the diagnosis of myofibroma.

Currently the patient is on third-year follow-up without any evidence of tumor recurrence.

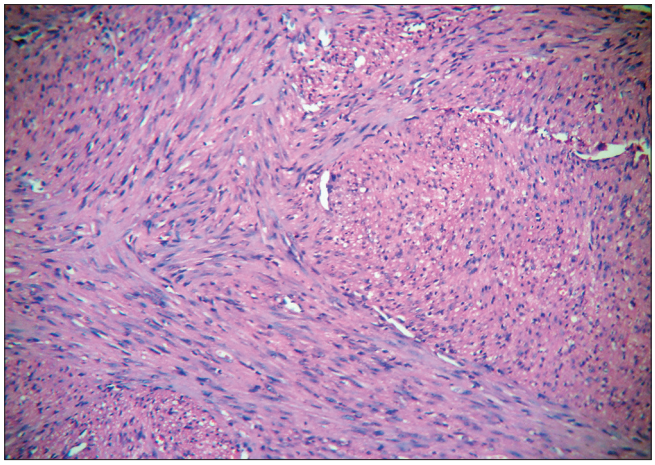

Figure 3: Photomicrograph showing biphasic nature of myofibroma with elongated spindle cells seen alternating with closely packed cells with small rounded nuclei and eosinophilic cytoplasm forming interlacing fascicles. (H and E, stain ×200)

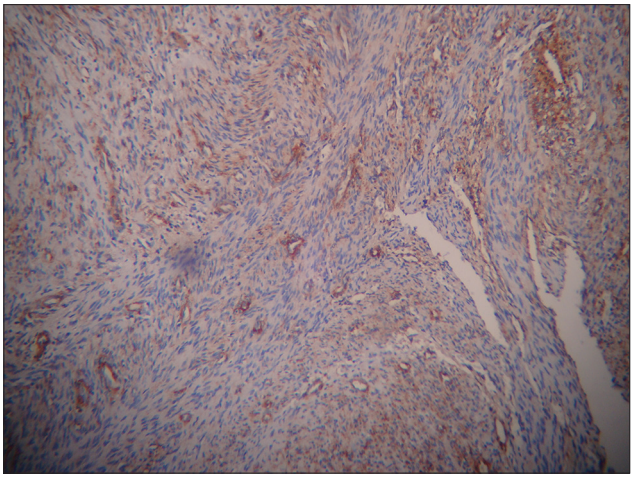

Figure 5: Strong immunoreactivity for αSMA in the tumor cells and hemangiopericytoma-like vascular pattern is seen. Note that the blood vessel walls are also positive (×100)

DISCUSSION

Myofibroma/myofibromatosis is a rare tumor presenting as solitary or multiple lesions with a predilection for soft tissues of the head and neck region. It is less common within the jaw bones. Myofibroma is thought to represent a benign proliferation of the myofibroblast, a cell with a phenotype of both fibroblast and smooth muscle cell, as demonstrated by immunohistochemical, histomorphologic, and ultrastructural studies.[4]

Central (intraosseous) myofibroma of mandible is rare. Allon *et al*.,[5] in 2007 reported four new cases of myofibroma of the mandible, in addition to the 19 other well-documented cases that had been reported until then. The 12 cases mentioned in AFIP review[6] were excluded from their analysis due to lack of sufficient data. Since then, only three additional cases of myofibroma of mandible have been reported,[7-9] making our case the 39th such report in the medical literature.

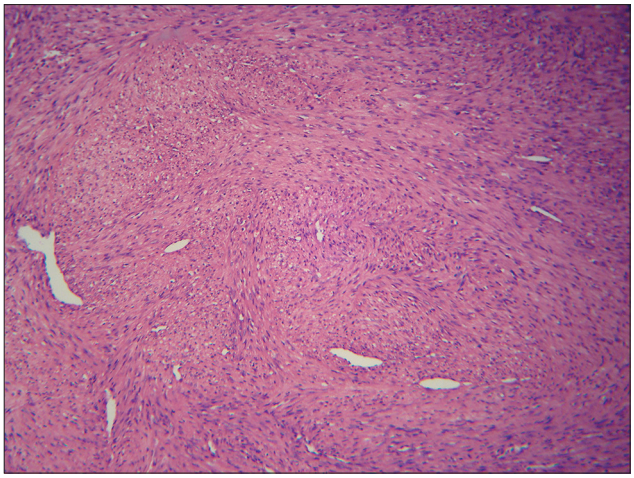

Figure 4: Hemangiopericytoma-like vascular pattern is seen. (H and E, stain ×100)

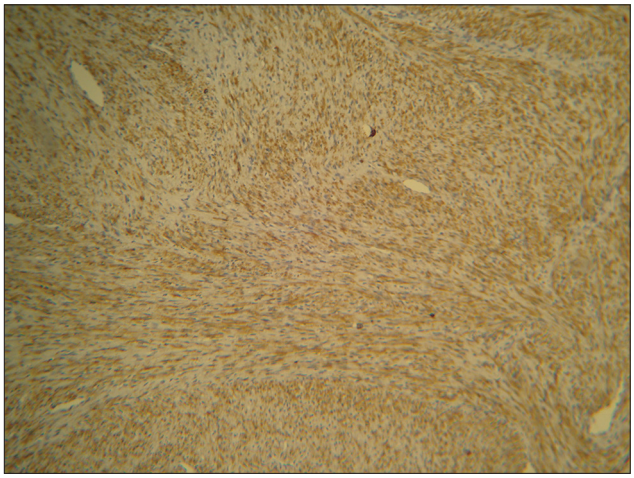

Figure 6: Photomicrograph demonstrating strong immunoreactivity for αSMA in the tumor cells showing a biphasic pattern. (×100)

Myofibroma of the mandible is commonly diagnosed in children in the first decade of life and shows a definite male predilection. Clinically, lesions present as an asymptomatic jaw swelling and rarely as a soft tissue mass when there is cortical plate perforation, as was the case in our patient.

Radiologically myofibromas are usually unilocular radiolucent lesions with well-defined borders. The present case also revealed a unilocular osteolytic lesion displacing the impacted 48 inferiorly. The most likely differential diagnoses include ameloblastoma (unicystic type), ameloblastic fibroma, and odontogenic keratocyst. Lesions less likely to be considered were the radiolucent varieties of the calcifying cystic odontogenic tumor and central odontogenic fibroma.[10] If myofibroma shows a multilocular appearance, differential diagnosis should be extended to include central hemangioma, aneurysmal bone cyst and ameloblastoma (solid type). When there is an ill-defined radiolucency in central myofibroma, the differential diagnosis should include aggressive lesions like desmoplastic fibroma and Ewing's sarcoma.[5]

Histopathologically, myofibroma is characterized by a nodular biphasic pattern. Lightly stained areas comprising fascicles of myofibroblasts with abundant extracellular matrix are seen. Cells are spindle to ovoid shaped with little observable pale cytoplasm. In addition, darkly stained areas consisting of smaller, densely packed, round to spindle-shaped myofibroblasts with intense eosinophilic cytoplasm associated with hemangiopericytoma-like vascular pattern are also noticed.[6,11,12] The alternating presence of these two patterns in histopathologic sections creates a micronodular "zoning" phenomenon distinct for myofibromas, which is well appreciated in the present case.

The histopathological differential diagnosis of myofibroma must include the tumors of muscle origin, neural origin and certain tumours like desmoplastic fibroma, fibromatosis, and low-grade fibrosarcoma. Lesions of neural origin can be excluded based on immunopositivity with S100, which is absent in myofibroma. Leiomyoma and leiomyosarcoma of bone are rare and can be distinguished by their reactivity for desmin.[13] Moreover, leiomyosarcomas possess considerably more cellular pleomorphism and higher mitotic rate. Myofibroma possesses neither the blunt ended cigar shape nuclei of leiyomyoma or leiomyosarcoma, nor are the cells arranged in long fascicles intersecting at right angles.

Differentiation from solitary fibrous tumor may also be difficult because of hemangiopericytoid appearance in both lesions. Solitary fibrous tumor is described as a pattern-less proliferation of spindle cells with alternating hypercellular and hypocellular areas rich in a dense keloid type of collagen.[14] It can be differentiated from myofibroma by its characteristic immunoreactivity for CD34 and CD99 which is negative in myofibroma.[15]

Desmoid-type fibromatosis has an aggressive behavior and morphologically desmoplastic fibroma (DF) of bone resembles myofibroma. The infiltrative and destructive growth pattern of DF and the absence of hemangiopericytoma-like vascular pattern can help in differentiation. DF shows less constant positivity to αSMA and HHF35 which are well established in myofibroma.[5,16] Fibromatosis has a more monomorphic growth pattern comprising long sweeping fascicles of spindle cells among abundant wavy collagen fibrils, which is not a feature of solitary myofibroma.

Fibrosarcomas of the bone can be differentiated from myofibroma by the presence of features like "Herring bone pattern," nuclear atypia, high mitotic activity including abnormal mitoses,[5,17] necrotic and hemorrhagic areas, etc.[11] These features are usually absent in myofibroma. Furthermore, fibrosarcomas do not display the zoning phenomenon of myofibroma.

Myofibroma and myofibrosarcoma show a significant degree of overlap in clinical and morphologic features as both have predilection for the head and neck region. Evaluation of desmin expression may be a diagnostic adjuvant where myofibroma stains negative and a significant portion of myofibrosarcoma stains positive for desmin.[10]

Treatment of myofibroma of the mandible is usually conservative excision. Very few cases need an aggressive surgical management like segmental jaw resection to remove extensive and destructive tumors.[5] Prognosis of the solitary adult lesions appears to be excellent with few recurrences of the oral lesions so far being recorded and these were cured by further local curettage.[10]

Though controversy exists regarding the histogenesis and nature of true myofibroblastic lesions, many tumors in routine surgical pathology show convincing features of myofibroblastic differentiation.[18] The cause of myofibroma is presently unknown. A number of authors have suggested that the tumors are inherited in an autosomal dominant[19] or autosomal recessive trait.[20] However, its low familial incidence suggests that there are probably factors other than genetics that play an important role in the etiology of this disease.

To summarize, myofibroma is a benign tumor that occurs in childhood and adolescents with a preference to mandible, when occurring in the jaw bones. Most myofibromas of mandible are slow growing, produce unilocular well-defined radiolucent lesions and have a tendency to expand and perforate the cortical plate. Histologically, it is similar to myofibroma elsewhere in the oral cavity producing a biphasic/zoning pattern and must be differentiated from certain benign and malignant spindle cell neoplasms. Local-wide surgical excision is the treatment of choice. The tumor has no tendency for recurrence and has an excellent prognosis. A correct and early diagnosis of myofibroma in central lesions involving mandible can help avoid aggressive surgical procedures.

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