Central myofibroma of the maxilla

Paramjeet Kaur, Rajat Chowalta, Jeevan Lata

Abstract

Myofibroma is a rare benign localized or generalized proliferation of myofibroblastic tissue occurring mostly in infants or children. In the oral region, most lesions occur in the mandible, lip, buccal mucosa, and tongue; however, the lesions arising in the maxilla are very rare. Myofibroma has an aggressive clinical presentation and is often treated aggressively because of an inappropriate diagnosis. A unique feature of central myofibroma of the jaws is the potential for teeth and other odontogenic structures to be involved by tumor. We report a case of myofibroma arising in the left side of the maxilla of a 12-year-old girl and describe the differential diagnosis from other spindle cell lesions of neural and smooth muscle origin. We treated the case using surgical excision under general anesthesia. Immunohistochemical staining was done for establishing the diagnosis since histopathological diagnosis with conventional staining could not distinguish myofibroma from spindle cell tumors.

Keywords: Central myofibroma, immunohistochemical staining, maxillary bone

Introduction

Solitary central myofibroma[1-3] is a rare neoplasm that typically arises in soft-tissue and subcutaneous sites in the head and neck, but rarely within bone. The lesion is thought to represent a benign proliferation of the "myofibroblast."

Foss and Ellis[4] analyzed 79 cases of myofibroma in the oral cavity and reported only four cases in maxilla. Montgomery *et al*.[5] analyzed nine cases of myofibroma of oral cavity and found no intraosseous maxillary tumor.

In the oral region, most lesions occur in the mandible, lip, buccal, mucosa, and tongue. However, the solitary myofibroma of maxilla is very rare.

Case Report

A 12-year-old female child reported to the Department of Oral and Maxillofacial Surgery of Punjab Government Dental College and Hospital with the chief complaint of painless swelling in the left upper front region of face since 8 months.

Extraorally, the face was asymmetrical with swelling present on left anterior maxillary region measuring 6 cm mediolaterally × 5 cm superioinferiorly. The swelling extended up to the lateral wall of the nose medially and infraorbital rim superiorly causing decrease in the size of left palpebral fissure [Figure 1]. The overlying skin was normal in color and texture and not adherent to the underlying tissue swelling was firm in consistency on palpation. There was no complaint of any paresthesia.

Intraorally, the swelling extended from left upper central incisor to first molar, buccopalatally. There was displacement of canine. The first and second premolars were clinically missing in the left maxillary region. The canine was slightly mobile, and all the involved teeth were vital. Overlying mucosa was smooth but slightly inflamed with no surface ulceration. The aspiration test was negative.

Panoramic radiograph showed well-defined radiolucency extending from canine to the first molar with distal tilting of canine and the first premolar and medial tilting of the first molar. Computed tomography (CT) revealed large expansile soft-tissue attenuation mass with relation to left maxilla with area of necrosis. The mass was causing thinning and erosion of walls of maxillary antrum and medially causing partial compression of nasal airway [Figure 2]. No significant

Department of Oral and Maxillofacial Surgery, Punjab Government Dental College and Hospital, Amritsar, Punjab, India

Correspondence: Dr. Rajat Chowalta,
Rajat Villa, H. No. 76, Shakti Nagar Jaunaji Road, Solan - 173 212, Himachal Pradesh, India.
E-mail: rajat.chowalta@gmail.com

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lymphadenopathy was seen. CT scan report suggested nonossifying fibroma.

On the basis of clinical and radiographic examination, it was provisionally diagnosed as an ossifying fibroma of maxilla. Ameloblastoma, keratocystic odontogenic tumor, odontogenic myxoma, and adenomatoid odontogenic tumor were kept as differential diagnosis. Considering the well-defined nature of lesion, excisional biopsy was planned.

On exposure, an encapsulated tumor mass was directly visualized because the alveolar and maxillary bone were destroyed. Since the canine and the first premolar were in close contact with the tumor, the whole of the tumor mass was easily shelled out from the surrounding tissues along with the two teeth and the specimen was sent for histopathological examination. The resected tumor was firm, encapsulated mass measuring 6 cm × 5 cm with well-defined borders and a whitish surface [Figure 3].

The histopathological report revealed that the lesion contains sweeping fascicles of spindle-shaped cells with whorling patterns at many places. Small slit-like vascular spaces (hemangiopericytomatous pattern) were seen with areas of calcification noted. No mitotic activity was seen. Immunohistochemical analysis revealed that the tumor cells were positive for vimentin and alpha-smooth muscle actin (α-SMA), and negative for Ki67 and S-100 and focal positivity for desmin. On the basis of strong positivity for vimentin and α-SMA, the tumor was immunohistologically diagnosed as a myofibroma of the maxilla. In our case, the complete surgical excision of the lesion was performed along with involved teeth. There is no report of recurrence until date.

Discussion

Myofibromas and myofibromatosis represent the mysterious group of lesions that were first described by Stout[6] as congenital multicentric fibroblastic proliferation. They were eventually characterized as phenotypically myofibroblastic tumors of infants and children by Chung and Enzinger[7] who noted the predominance of solitary tumors compared with tumors of multicentric origin.

Williams and Schrum,[8] first, classified these lesions as congenital fibrosarcomas. The patients were newborns and infants who presented with multiple nodular lesions of the skin, subcutaneous tissue, muscle, bone, and viscera. These visceral lesions adversely affect the prognosis.

Kauffman and Stout[9] subsequently subclassified congenital fibromatosis into multiple and generalized forms. However, Jones *et al*.[2] in accordance with Smith *et al*.[10] preferred the term myofibroma when describing a solitary neoplasm of this type.

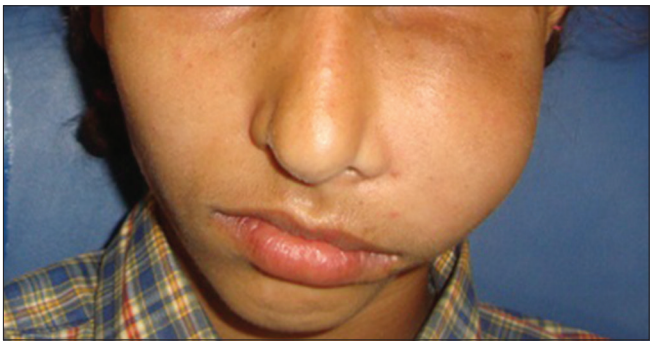

Figure 1: Extraoral swelling extending up to the lateral wall of nose medially and infraorbital rim superiorly causing decrease in the size of left palpebral fissure

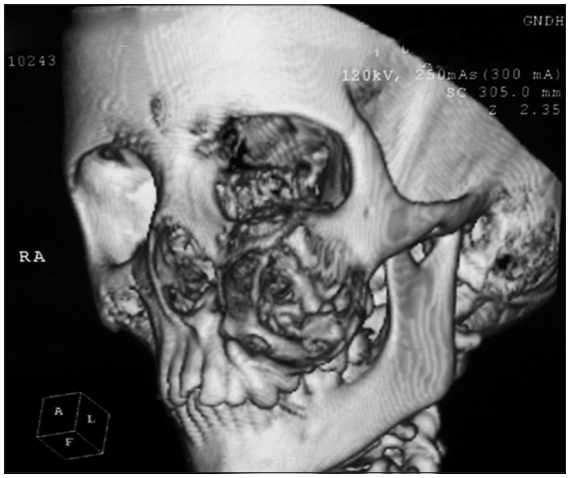

Figure 2: Three-dimensional computed tomography face showing the extent of involvement of lesion

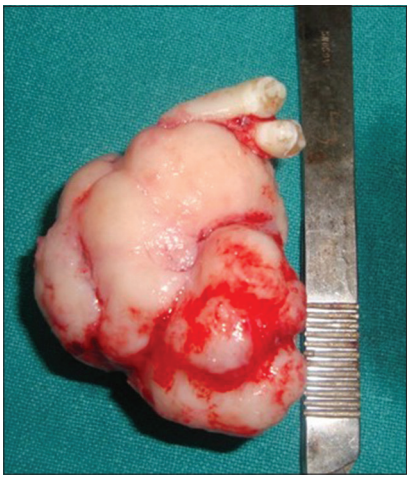

Figure 3: Firm, encapsulated mass measuring 6 cm × 5 cm with well-defined borders and a whitish surface

The solitary myofibroma is a benign lesion found in the superficial soft-tissues and recently has been recognized as separate entity from the more aggressive deeper multiple lesions of infantile myofibromatosis. Soft-tissue lesions occur predominantly in the head and neck, including mouth, and

the diagnostic criteria are described for other body sites. Intraosseous lesions are much less common but have a marked predilection for the skull and the mandible.

The etiology of myofibromas is presently unknown. Many authors have worked on this and suggested that the tumors are inherited as an autosomal dominant[11] or an autosomal recessive[12] trait. As the lesion develops from the benign proliferation of myofibroblasts, these cells are thought to play an important role in wound healing.[13] This suggests that a previous history of trauma may contribute to the development of these lesions.

Clinically, the lesions are more often seen in infants although the age range is wide. The presenting symptoms depend on site of involvement. In most of the cases, periosteal expansion may be slight or it may be a coincidental finding on the radiographic examination. Erupted and unerupted teeth may be displaced. Radiographically, the lesions appear benign and are often assumed to be odontogenic tumor or cyst,[14] because they are sharply defined radiolucencies with a variably thick sclerotic rim.

Histologically, the lesions contain two distinctive patterns seen in soft-tissue lesions. Sweeping fascicles or whorled spindle cells lie in a variably collagenous matrix. The more fibrous areas stand out as pale-staining poorly demarcated patternless zones which at low power resemble scar or chondroid. They contain larger elongate spindle- or strap-like cells often with an eosinophilic cytoplasm. The second pattern is densely cellular and comprises rounded or ovoid cells arranged with no particular architecture but containing cleft-like vascular spaces. The vascular spaces are often described as hemangiopericytomatous, but large stag's horn formations are unusual in intraosseous lesions and clefts are smaller and less numerous than in soft-tissue lesions. Occasional mitoses may be present in the cellular areas, and calcification may occur in the scar-like areas. The zoning phenomenon, which is useful in the diagnosis of soft-tissue lesions, is often not apparent in intraosseous lesions.

Due to these findings, this disease is often misdiagnosed as benign and malignant spindle cell lesions of nerve tissue or smooth muscle origin (leiomyoma).[15,16]

Hence, immunohistochemical staining[17] is a useful tool to identify the nature of neoplastic cells and to reach an accurate diagnosis. Vimentin is the most widely expressed intermediate filament protein thought to be involved in structural processes. This protein is a general marker of cells originating in the mesenchyme. α-SMA expression is the most used marker for myofibroblast identification.

Neural lesions can be excluded because of their immunoreactivity with S-100, which are absent in myofibroma. Thus, understanding the clinical and pathological characteristics of myofibroma is very important to establish the correct diagnosis and avoid unnecessary treatment modalities.

Immunohistochemistry helps distinguish myofibroma from fibrosarcoma, the former is positive for vimentin, α-SMA and negative for desmin and S-100, whereas the latter is negative for α-SMA, also fibrosarcoma of the bones can be differentiated from myofibroma by the presence of a "herring-bone" pattern, nuclear atypia, and high mitotic counts including abnormal mitoses.

Other possible misdiagnoses include leiomyoma,[15,16] schwannoma, fibrous histiocytoma, fibroma, hemangiopericytoma, and nodular fasciitis.[18] Nodular fasciitis gives a short preoperative duration of not more than 1–2 months in most, but not all, cases with tenderness. Extravasated red cells, chronic inflammatory cells, and multinucleated osteoclast-like giant cells are other frequently identified features. Nodular fasciitis also differs from myofibroma in its tendency to be mitotically active. Schwannoma, fibrous histiocytoma, fibroma, and hemangiopericytoma are negative for α-SMA whereas leiomyoma is positive for desmin. Such immunohistological characteristics allow these tumors to be distinguished from myofibroma. Because the tumor in our patient was positive for vimentin and α-SMA, it was diagnosed as a myofibroma.

Curettage is generally curative as the lesions shell out readily. In case of large lesions, resection with 0.5 cm is advisable and occasional recurrence is to be expected.

Conclusion

Central myofibroma is a typical tumor of child-hood and adolescents in the gnathic region. Solitary myofibromas of the head and neck region, such as other sites, are biologically inert and show very little or no recurrence following excision. However, because of complex anatomy and limitations in the head and neck, especially the gnathic region, a dramatic radiographic and clinicopathologic presentation is not uncommon, making the suspicion for malignancy high in such cases. Thus, accurate diagnosis via pathological findings including immunohistochemistry is essential to avoid wrong diagnosis and the consequent unnecessary pervasive treatment therapies.

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Conflicts of interest
There are no conflicts of interest.

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