

Benign Spindel Cell Neoplasm: A Central Myofibroma of Maxilla in Male Child of Seven Years of Age: A Case Report



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Myofibroma is a rare benign spindle cell neoplasm which occurs predominantly in infants and young children. It can occur as a solitary mass or as a multicentric lesion, consisting of myofibroblasts. In oral cavity, the sites most commonly involved are the mandible, tongue, lips and buccal mucosa however, it rarely involves the maxilla. It has aggressive clinical presentation mimicking malignancies. We report a case of a male child seven years of age presented with intraoral swelling on the left palatal aspect of the maxilla that extended from deciduous upper left second molar to the distal of upper permanent left first molar causing the mobility of these teeth. Radiologically, the lesion was osteolytic causing marked resorption of alveolar bone and displacement of permanent maxillary left second molar. A differential diagnosis of peripheral giant cell granuloma, central giant cell granuloma, fibroma, aggressive fibromatosis, nodular fasciitis, peripheral ossifying fibroma and spindle cell neoplasm was made. After the excluding malignant and vascular involvement, the surgical excision of the lesion was planned and was carried out under general anesthesia. Histologically, benign proliferation of spindle cells was seen giving the diagnosis of benign spindle cell neoplasm. Reports also showed positive immunoreactivity with vimentin and α SMA while non-responsive to desmin, validates the diagnosis of myofibroma.

KEYWORDS: spindle cell neoplasm; myofibroma; maxilla

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INTRODUCTION

Myofibroma was first described in 1954 by Stout as "congenital generalized fibromatosis"¹ which was renamed as "infantile fibromatosis" by Chung et al. in 1981.¹ However, later in 1989 Smith et al introduced the term "myofibroma" for these lesions.¹ Afterwards, the terms "myofibromatosis" and "myofibroma" were adopted by the World Health Organization.² It can occur as solitary or multicentric lesion.³ The presence of solitary lesion is uncommon in oral and maxillofacial region and the prevalence is not more than two percent of the total

cases reported. Oral and perioral regions are commonly involved. However, intraosseous lesion of the jaw most commonly accounts for mandible and is rarely found in maxilla.³

Myofibroma occurs most commonly within the first ten years of life and 90% of these cases usually present before 2 years of age.⁴

The exact etiology of this condition is unknown with most reported cases suggestive of its sporadic nature; however, some cases reported its familial pattern of inheritance.⁴

Clinically Intraosseous myofibroma usually shows swelling of the jaws and sometimes when perforated can be presented as soft tissue mass. The distinctive characteristic of central myofibroma of the jaws is that it includes the teeth that displays clinical or radiographic picture indicative of odontogenic or nonodontogenic lesions.⁵

In this study, a central myofibroma of maxilla in a male child of seven years of age is presented, which is a very rare clinical entity.

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CASE REPORT

A male child of seven years of age presented to the outpatient department of pediatric dentistry, children hospital, PIMS, with the complaint of swelling in upper left jaw extending to the palate for two months that was slowly increasing in size and was causing eating difficulties



Figure 1

(Figure 1). There was history of trauma in the involved site two months back. There was no medical history and the family history was also not significant. Oral hygiene of the patient was compromised. The swelling was asymptomatic with no pain and bleeding. However, the lesion occasionally bled slightly on manipulation. An informed and written consent was also taken from the patient or the attendant.

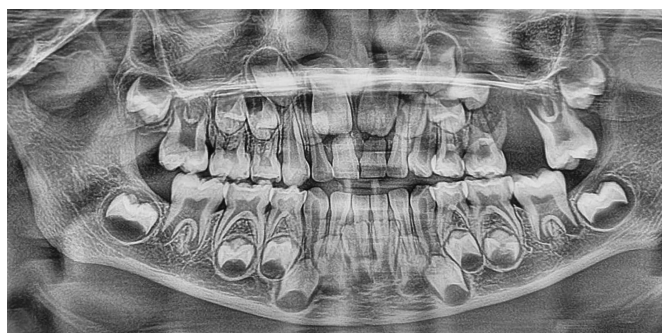
On extra-oral examination, there was no facial swelling or asymmetry with no associated lymphadenopathy. On intraoral examination there was a 3×3cm pedunculated, pinkish soft tissue mass on palatal aspect of the attached gingiva extending from upper primary left second molar to the distal of permanent left first molar causing the mobility of these teeth (Figure 2). The mass



Figure 2

had a firm consistency on palpation with no bruit and central ulceration thus excluding the vascular involvement. Intraoral periapical radiograph and orthopantomogram revealed marked resorption of alveolar bone along with distal drifting of permanent maxillary left second molar. (Figure:3).

Figure 3



A provisional diagnosis of central giant cell granuloma was made. Peripheral giant cell granuloma, central giant cell granuloma, fibroma, aggressive fibromatosis, nodular fasciitis, peripheral ossifying fibroma and spindle cell neoplasm were kept as differential diagnosis. After performing all baseline blood investigations, and anesthetist and maxillofacial surgeon consultation, excisional biopsy of the lesion was planned under general anesthesia excluding any other underline co-morbidities. Since upper

primary left second molar and permanent left first molar was in close approximation to the lesion so, the teeth were extracted along the lesion (Figure:4). Following extraction of the teeth blunt dissection was done to remove all the remnants of the lesion. Primary closure was done with vicryl 3/0 followed by the placement of an absorbable hemostat, surgicel (Ethicon, 10.2 cm×20.3cm) to control bleeding and periodontal dressing Coe Pak (Gc America) to facilitate healing (Figure:5). The specimen was

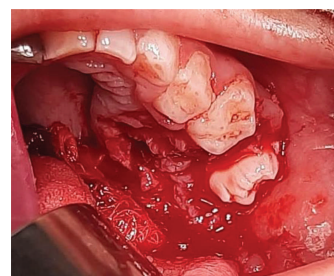


Figure 4

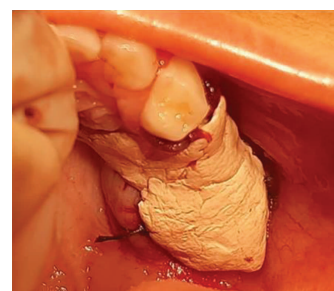


Figure 5

immersed in 10% formalin and was submitted for histopathologic examination. Histopathological studies revealed the benign proliferation of spindle cells arranged in streaming fascicles running in all directions giving the diagnosis of benign spindle cell neoplasm (Figure:6).

Figure 6

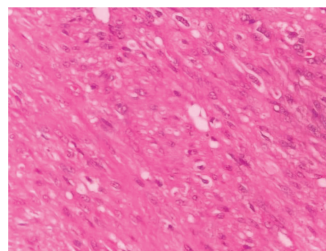


Figure7

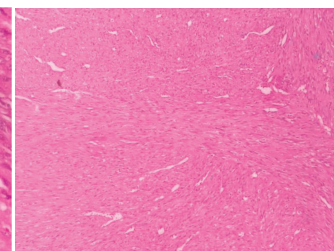
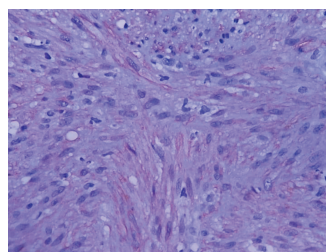


Figure 8



Immunohistochemical studies were done for the definitive diagnosis and the specimen showed positivity for α SMA and vimentin and negativity for desmin which was suggestive of myofibroma of the maxilla (Figure:7). The patient is currently undergoing routine follow-up (figure:8,9).

Figure:9 (Follow up after 4 months)



DISCUSSION

Myofibroma or myofibramatosis is a rare neoplasm of mesenchymal origin which is benign in nature, and consists of myofibroblasts.. It can be present as single lesion (myofibroma) or multiple (myofibramatosis) lesions. Soft tissues in the head and neck region are usually involved and are rarely found in the jaws, with only few cases reported in literature.⁵ However, when found in jaws, mandible shows greater predilection as compare to maxilla.⁶ In the present case the patient presented with the solitary myofibroma of the maxilla.

The etiology of these lesions is unclear. Some authors suggested that they are transmitted as an autosomal dominant or recessive characteristic while others suggested that trauma can be an important causative factor. They were of the view that as benign proliferation of myofibroblasts plays an important role in the wound healing then trauma to the tissues can be the leading cause in the formation of such lesions.⁷ The etiology in the present case can be the trauma which have resulted in the proliferation of myofibroblasts. Myofibromas are most commonly found in early age with slight male predilection.⁸ Clinically the lesion presents as hard, protuberant swelling with no symptoms resulting in bony expansion of the jaws and disturbing the facial symmetry.⁹ In our case a hard, pedunculated, movable mass was noted on the palatal aspect of left posterior maxillary region in a male child seven years of age which is suggestive of the clinical findings of myofibroma as mentioned in various reported studies.

Myofibromas can be present as unilocular or multilocular radiolucency encircled by a well-defined border on radiograph.

Some case reports presented thinning of the cortical plate along with the displacement of involved teeth.¹⁰ The present case also shown a unilocular radiolucency, with excessive bone resorption resulting in mobility of primary maxillary left second molar and permanent maxillary left first molar and displacement of permanent maxillary left second molar.

Histologically the lesion consists of light and dark stained regions. Spindle cells contributes to the light area of the lesion which contains eosinophilic cytoplasm and tapered shaped nuclei at the periphery of the lesion whereas, the dark areas are composed of round or spindle shaped cells which are distributed centrally. These cells contains basophilic nuclei with eosinophilic cytoplasm and hazy cell borders. The light and dark stained regions produces zoning pattern which is the characteristic of soft tissue lesions and are not found in intra osseous lesions.¹¹

These features are often misleading and the disease can be wrongly diagnosed as benign or malignant spindle cell lesions of nerve or muscle origin (leiomyoma).^{7,11} Immunohistochemical staining is the key to the accurate diagnosis. It aids to differentiate between myofibroma and fibrosarcoma. In case of myofibroma vimentin, α -SMA will be positive whereas desmin and S-100 will be negative, while in fibrosarcoma α -SMA will be negative. Certain distinctive characteristics such as abnormal mitoses, atypical nuclear features and "herring bone" phenomenon also helps to distinguish fibrosarcoma from myofibroma.¹² The present case showed positivity for α -SMA and vimentin which was suggestive of myofibroma.

Treatment involves complete surgical excision. Local recurrence has been found to be 7% to 31% in cases of myofibramatosis. Prognosis is quite good in case of solitary lesions however, it can be aggressive and fatal in case of multicentric lesions.^{9,12,13}

CONCLUSION

Central myofibroma is a benign tumor commonly present in young age. Mandible is commonly involved. The novelty in our case is that it showed a unique occurrence and presentation of myofibroma that originates from the maxilla. As myofibroma of the jaws can be very aggressive causing bone resorption and displacement of the permanent teeth sometimes mimicking malignancies so, thorough clinical and pathological findings are important tool along with the immunohistochemical studies to eliminate the risk of misdiagnosis and unnecessary invasive treatment modalities.

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ROLE AND CONTRIBUTION OF AUTHORS:

H.N data collection and drafted the manuscript. **A.K.Z** Conception and design of the study, Collection of data and assembly and article writing **M.I** data collection and drafted the manuscripts **S.M** and **A.I** data collection, drafted the manuscript and performed literature search.

CONFLICT OF INTERESTS

The authors declare that they have no conflicts of interest in relation to the publication of this article.

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None to declare

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