

Intraosseous infantile myofibroma of the mandible

Access this article online

Website:
www.amsjournal.com

DOI:
10.4103/2231-0746.83151

Quick Response Code:



Natarajan Nirvikalpa, Vinod Narayanan

Department of Oral and Maxillofacial Surgery, Saveetha Dental College and Hospital,
Chennai, Tamil Nadu, India

Address for correspondence:

Dr. Natarajan Nirvikalpa, Department of Oral and Maxillofacial Surgery,
Saveetha Dental College and Hospital, Chennai – 600 077, Tamil Nadu, India.

E-mail: nirvikalpa9@hotmail.com

ABSTRACT

The myofibroma, especially the intraosseous variety, is an uncommon benign tumor that occurs primarily in children younger than 3 years of age. Since 1966, less than 40 cases of solitary myofibromas of the mandible have been reported in the literature. Myofibroblasts and spindle cells are predominantly found in this benign lesion. These cells are also commonly found in many lesions due to which great difficulty can be encountered in the diagnosis. A rare case of the intraosseous variety of an infantile myofibroma of the mandible diagnosed in a 10-month-old child has been reported. The tumor was completely excised via an intraoral approach and no recurrence has been noted 15 months postoperatively. A brief review of the differential diagnosis of this lesion in terms of its clinicopathologic, histologic, and immunohistochemical features is also discussed.

Keywords: Infantile myofibroma, intraosseous, mandible

INTRODUCTION

Myofibroblasts are spindle-shaped cells with features of both fibroblasts and smooth muscle cells. Infantile myofibromatosis is a benign, self-limiting (localized or generalized) growth of probable hamartomatous origin consisting of cells that show the characteristics of myofibroblasts and/or smooth muscle-like cells.^[1,2]

Myofibroma is a rare but well-recognized condition that was first described by Stout in 1954 as “congenital generalized fibromatosis.”^[3]

In 1981, Chung and Enzinger proposed the term infantile myofibromatosis to replace Stout’s terminology for these lesions, because of the resemblance between certain clinical features of congenital generalized fibromatosis and localized fibromatosis and in view of the occurrence of the lesion specifically in both newborns and infants.^[4]

On the rare occasion when it appears in the oral cavity, it usually presents as a locally invasive, hard painless mass. The classical histopathological picture is that of a well-circumscribed neoplasm with a biphasic pattern consisting of fascicles of spindle-shaped

cells admixed with areas that demonstrate a hemangiopericytoma-like pattern.^[5]

This article intends to describe a case of this rare lesion and analyze the differential diagnosis of this lesion in terms of its clinicopathologic, histologic, and immunohistochemical features.

CASE REPORT

A 10-month-old boy was referred to Department of Maxillofacial Surgery with a painless swelling in the mandibular left posterior region that had been present for 5 months. The lesion had gradually increased in size. Medical and familial histories were noncontributory. There was no history of trauma or spontaneous bleeding associated with the lesion. No lymph nodes were palpable in relation to the lesion. An extraoral examination showed a diffuse swelling, firm in consistency, approximately 3 × 3 cm, extending from the angle of the mandible to the middle of the ramus and superoinferiorly from 2 cm below the malar prominence to the inferior border of the mandible [Figure 1]. There was no extraoral discharging sinus and the skin over the lesion appeared normal. Intraoral examination showed an expansile mass involving buccal and lingual cortices in the retromolar and molar region.

The two-dimensional (2D) CT scans [Figures 2 and 3] showed a lesion approximately 2.7×2.1 cm in the posterior mandible on the left side with broad expansion of the buccal and lingual cortices and well-defined borders. The mass showed central soft tissue attenuation and well-defined borders. Bony erosion at the margins was also seen. Further analysis of the 3D CT confirmed the extent of the lesion and also revealed areas of bone destruction in the buccal and lingual cortices. It also showed infringement of the lesion up till the coronoid and condylar process.

Incisional biopsy established the diagnosis of myofibroma, and complete surgical excision along with a peripheral osteotomy was planned.

Under general anesthesia, nasotracheal intubation was done. After infiltrating with local anesthesia, the mucoperiosteal flap was raised and the lesion was exposed. The lesion was excised and a margin of normal bone of 1 cm, surrounding the lesion, was also removed. The inferior alveolar nerve was found to be displaced and was not damaged during the resection. The mass was removed in fragments from the bony crypt and was found to be unencapsulated. Thorough curettage of the bony crypt was carried out; gelfoam was used to fill the defect [Figure 4]. Postoperative recovery was uneventful. A 15-month follow-up period showed no signs of recurrence. Pre- and postoperative assessment of the inferior alveolar nerve could not



Figure 1: 10-month-old infant showing a diffuse swelling in the left mandibular posterior region

be carried out accurately due to the age of the child.

The excised mass was sent for histopathological analysis. The macroscopic presentation was that of multiple fragments of greyish white soft tissue, the largest measuring $2.5 \times 1 \times 0.8$ cm and the smallest measuring $8 \times 3 \times 2$ mm. The cut surface had a firm white appearance. Microscopic examination confirmed the mass as myofibroma and the histologic picture revealed the typical biphasic pattern with spindle cells, which were arranged in fascicles and bundles. The nuclei were spindle shaped with bright eosinophilic cytoplasm set in hyalinized stroma with a faint basophilic appearance. One section showed an area of hyalinization. Focally congested blood vessels and a cluster of foreign body-type multinucleate giant cells were seen. Fragments of bone were also seen on one aspect. A concomitant characteristic trait of a hemangiopericytoid pattern, i.e., a combination of smaller cells and slit-like vascular spaces, was prominent. The mitotic activity was absent.

Immunohistochemical analysis confirmed the diagnosis. The specimen tested positive for smooth muscle actin and vimentin [Figures 5 and 6]. Immunoreactivity for cytokeratin, desmin, and

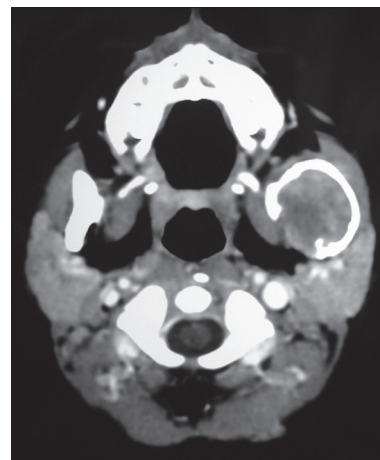


Figure 2: Axial CT demonstrating a well-defined lesion with the expansion of buccal and lingual cortical plates

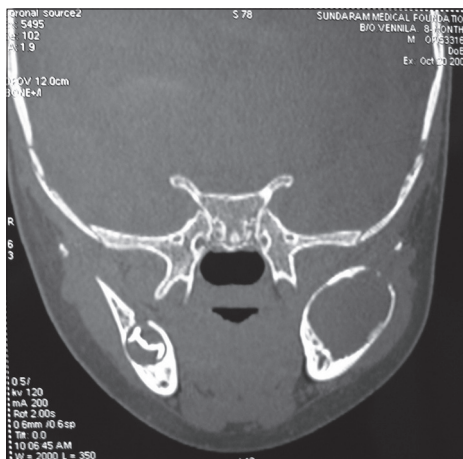


Figure 3: Coronal CT showing the expansion of buccal and lingual cortical plates and area of perforation of the buccal cortex



Figure 4: Intraoperative view after the excision of the mass and peripheral osteotomy of the posterior mandible

S-100 protein were negative.

DISCUSSION

To the best of the authors' knowledge, less than 40 cases of solitary myofibroma of the mandible have been reported so far in the literature.^[6]

Within the broad spectrum of recognized myofibroblastic conditions, myofibromas are best characterized as benign neoplasms, with the caveat that they sometimes exhibit features of both developmental lesions, such as congenital presentation and possible inheritable transmission, and of reactive conditions such as spontaneous remission and limited growth potential.^[7]

Myofibroma is considered to be a tumor afflicting both adults and children. Chung and Enzinger^[4] reported a series of 61 cases of infantile fibromatosis (45 solitary cases and 16 multiple cases) with a frequency of 32.8% in the head and neck region. The solitary form was more common in males (69%), primarily affecting tissues in the head, neck, and trunk. The multicentric form was more common in females (63%), being found in the soft tissues, skeleton, and viscera.

Myofibromas are preferentially known to occur in the head and neck region, trunk, and extremities, but rarely within the bone.^[8] When present intraosseously, a myofibroma appears as a slowly enlarging, asymptomatic, and expansile mass. Although the myofibroma is a benign neoplasm, it has a variable biologic behavior ranging from very mild to moderate degree of invasiveness. The tumor grows by infiltration and may produce cortical expansion, tooth movement, and root resorption. Radiographically, it resembles a cystic cavity. Enzinger and Weiss have reported the radiographic appearance of the lesion as circumscribed lytic areas with marginal sclerosis and without the penetration of the cortex.^[9] However, our case did show perforation of the buccal and lingual cortices.

Histopathology is probably the most variable aspect of this tumor. Histologically, myofibromas need to be differentiated mainly from leiomyomas, fibrosarcoma, nodular fasciitis, and hemangiopericytomas. Table 1 depicts the characteristic differentiating features of these lesions.

The presence of a zoning phenomenon specifically distinguishes myofibromatosis from other lesions, regardless of the fact that the cells in these lesions also exhibit immunoreactivity to smooth muscle actin.^[10] However, no specific histologic feature could be used to distinguish between tumors in adults and those in

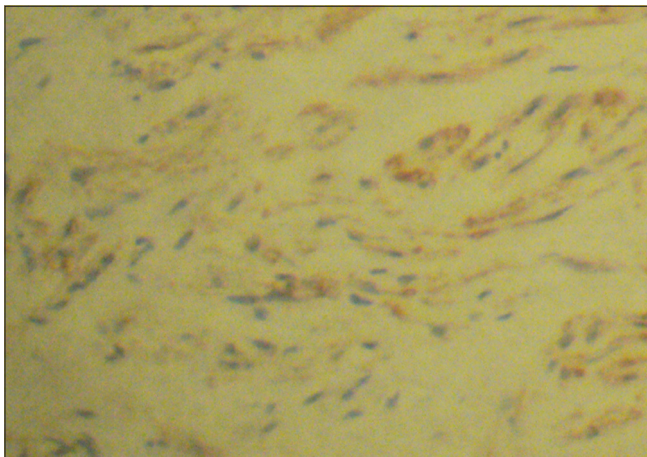


Figure 5: Photomicrograph of the SMA stain showing irregularly arranged fascicles of spindle-shaped cells with blunt-ended nuclei (40x)

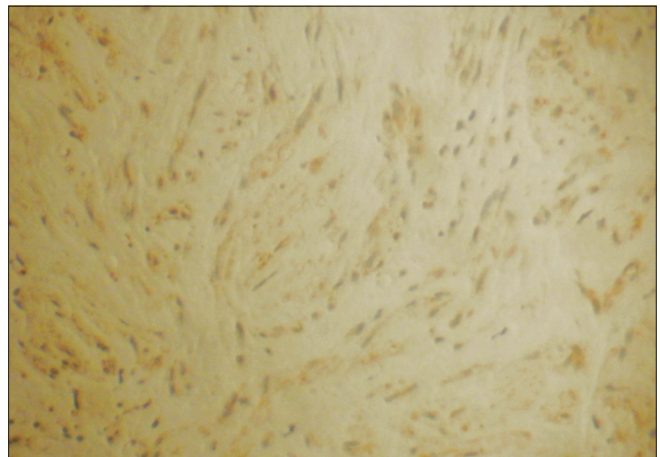


Figure 6: Photomicrograph of the vimentin stain of the lesion showing strong immunoreactions of the myofibroblasts (40x)

Table 1: Differential diagnosis of myofibroma with other jaw lesions showing myofibroblastic differentiation						
Lesion	Histology	SMA	Vimentin	Desmin	S100	Keratin
Myofibroma	Biphasic appearance of alternating spindle-shaped fascicular and cellular areas, circumscribed mass, ^[4] tapered nuclei ^[13]	Pos	Pos	Neg	Neg	Neg
Myofibrosarcoma	Diffuse infiltrative growth, herring bone fascicular growth, nuclear atypia ^[11]	Pos	Neg	Pos	Neg	Neg
Leiomyoma	Blunt-ended, cigar-shaped nuclei, cells arranged into long fascicles intersecting at right angles ^[11,13]	Pos	Neg	Pos	Neg	Neg
Leiomyosarcoma	Features of leiomyoma along with dysplasia ^[4,10]	Pos	Neg	Pos	Neg	Neg
Juvenile fibromatosis	Monophasic growth pattern of long sweeping fascicles of spindle cells among abundant wavy collagen fibrils. ^[11] Infiltrative growth ^[7]	Neg	Pos	Neg	Neg	Neg
Nodular Fasciitis	Feathery fascicles, ganglion-like cells and mucoid background. ^[12] Mitotically active. ^[13] Extravasated erythrocytes and lymphocytes ^[4]	Neg	Neg	Pos	Neg	Neg
Hemangiopericytoma	No peripheral mature area ^[10]	Neg	Pos	Neg	Neg	Neg

Neg = Negative; Pos = Positive

pediatric patients.^[11]

The distinction between other lesions especially fibromatoses and fibrosarcomas is important as these are more prone toward recurrence and metastatic change whereas myofibromas are usually cured by local excision. Stoolweg and Muller believe in a conservative approach as they observed a 7% recurrence rate for infantile myofibromatosis.^[8]

Matthews *et al.* have advocated that intraosseous jaw lesions that show predominantly fibroblastic or myofibroblastic cell population should initially be treated by thorough curettage or further wide excision where extension into soft tissue is identified.^[2]

CONCLUSION

Due to the rare and slow-growing nature of the tumor and follow-up often lost or only minimal; few reports of long-term outcomes or its biological behavior are available.

Factors to be borne in mind while deciding upon the selection of a surgical approach include the familial history of similar lesions, presence of lesions in other parts of the body, involvement of one or both cortical plates of the mandible, and extraosseous extension of the tumor into soft tissue. In our opinion, as myofibromas are unencapsulated tumors, treatment should include complete excision along with a border of clinically normal bone. A margin of about 1 cm is appropriate.

The ubiquitous nature of the myofibroblast and the benign, infiltrative nature of the tumor necessitate the achievement of a correct diagnosis. This is important for the administration of adequate treatment, that is, an appropriate resection with

accurate tumor-free margins.

REFERENCES

1. Dehner L. Tumors of fibrous tissue origin in children. *Cancer* 1976;38:888.
2. Matthews SM, Tabor MW, Thompson SH, Gross PD. Infantile Myofibromatosis of the Mandible. *J Oral Maxillofac Surg* 1990;48:884-9.
3. Stout AP. Juvenile fibromatosis. *Cancer* 1954;7:953.
4. Chung EB, Enzinger FM. Infantile myofibromatosis. *Cancer* 1981;48:1807.
5. Jones AC, Freedman PD, Kerpel SM. Oral myofibromas: A report of 13 cases and review of the literature. *J Oral Maxillofac Surg* 1994;52:870-9.
6. Allon I, Vered M, Buchner A, Dayan D. Central (intraosseous) myofibroma of the mandible: Clinical, radiologic, and histopathologic features of a rare lesion. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2007;103:e45-53.
7. Gold L. Infantile myofibromatosis of the mandible. *J Oral Maxillofac Surg* 1990;48:880-91.
8. Slootweg PJ, Muller H. Localized infantile myofibromatosis: Report of a case originating in the mandible. *J Maxillofac Surg* 1984;12:86-9.
9. Enzinger FM, Weiss SW. *Soft Tissue Tumors*. 3rd ed. St Louis, MO: Mosby; 1995. p. 238-45.
10. Liu CJ, Chang KW. "Infantile" myofibroma of the oral cavity: Report of case. *J Oral Maxillofac Surg* 2001;59:471-2.
11. Foss RD. Myofibromas and myofibromatosis of the oral region: A clinicopathologic analysis of 79 cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2000;89:57-65.
12. Montgomery EA, Meis JM. Nodular Fasciitis, its morphological spectrum and histological profile. *Am J Surg Pathol* 1991;15:942-8.
13. Montgomery E, Speight PM, Fischer C. Myofibromas presenting in the oral cavity: A series of 9 cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2000;89:343-8.

Cite this article as: Nirvikalpa N, Narayanan V. Intraosseous infantile myofibroma of the mandible. *Ann Maxillofac Surg* 2011;1:87-90.

Source of Support: Nil, **Conflict of Interest:** None declared.