

# Infantile Central Myofibroma of the Mandible: A case report

## Case Report

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## ABSTRACT

Oral myofibroma is an uncommon benign tumor that has great tendency to involve oral soft tissues with bony involvement remains a rarity. Most myofibromas exhibit a benign slow clinical course however intra-osseous lesions could be mistaken with many aggressive tumors which necessitate accurate diagnosis. The aim of this report is to shed light on a rare case of infantile central myofibroma of the mandible. Case report: A 6-month-old girl presented with an expansible painless left mandibular swelling. Radiographic examination revealed a well-defined, unilocular, radiolucent lesion with cortical erosions. Histopathological examination showed a highly cellular, encapsulated mass comprised of spindle-shaped and smaller round or polygonal cells in a vague biphasic pattern. Immunohistochemically, tumor cells showed strong cytoplasmic staining for  $\alpha$ -SMA, weak Ki-67 and negative desmin, CD34 and S-100 protein. Accordingly, conservative surgical excision with bone curettage was performed. Postoperative follow-up revealed excellent healing with no evidence of recurrence.

**Key Words:** Intra-osseous, mandible, myofibroblast, oral myofibroma,  $\alpha$ -SMA.

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## INTRODUCTION

Myofibroma is an uncommon benign tumor of myofibroblastic origin that sometimes becomes a challenging diagnosis<sup>[1]</sup>. Myofibromas mostly occurred as solitary or multiple lesions in skin, bone, lung and gastrointestinal tract of infants<sup>[2, 3]</sup>.

Myofibromas of the oral region has a marked propensity to involve soft tissues primarily tongue, buccal mucosa, palate and floor of the mouth<sup>[4, 5]</sup>. Intraosseous involvement is rarely encountered with the mandible being more commonly affected than maxilla<sup>[6]</sup>. Clinically, oral myofibroma usually presents as an asymptomatic pink mass or a swelling that interferes with mastication<sup>[1]</sup>. Central lesions may follow a more aggressive course displacing teeth or developing tooth follicles, causing tooth mobility or may evolve into an exophytic oral mass secondary to cortical perforation<sup>[1, 7- 9]</sup>.

Most myofibromas follow a benign clinical course with slow and limited growth<sup>[6]</sup>. Conservative surgical excision was determined to be the treatment of choice with only rare recurrences<sup>[4, 10]</sup>.

The purpose of this report is to better understand the clinical and histopathological characteristics of infantile myofibroma which are crucial for establishing correct diagnosis and avoiding unnecessary extensive therapies.

## CASE REPORT

A 6-month-old girl was referred to Oral and Maxillofacial Department, Faculty of Dentistry, Tanta

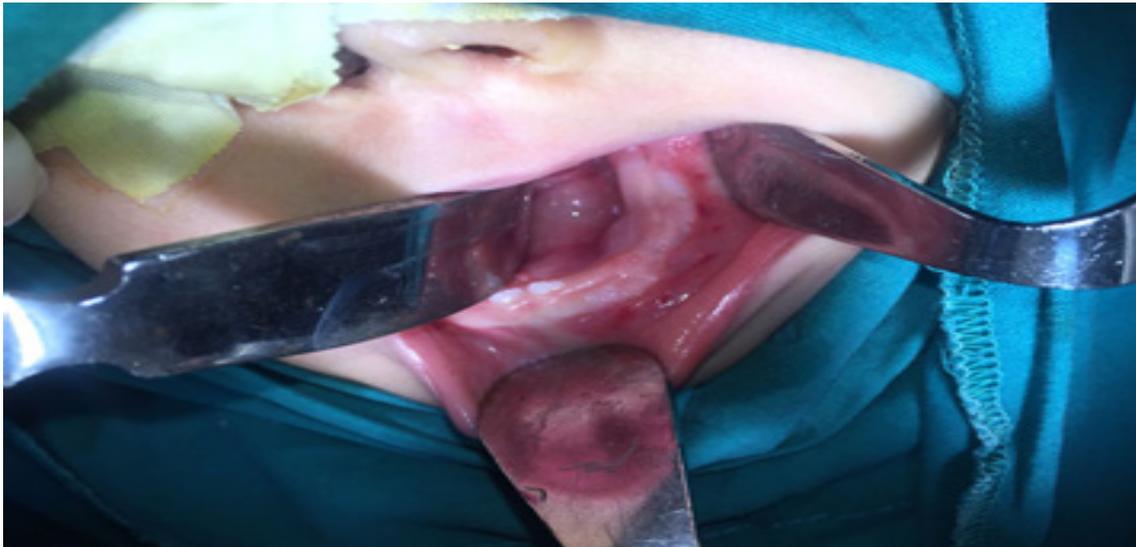
University, Egypt for evaluation of a painless gingival swelling at the left mandible which was first noticed by her parents 3 weeks ago and gradually increased in size. Her medical history was irrelevant. Furthermore, no history of trauma was associated with the lesion.

Before any procedure informed consent was obtained from her parents according to the guidelines on human research of the research ethics committee of faculty of dentistry, Tanta University. The proposed operation or investigations were explained in simple language which could be understood by the patient relatives. The more common complications were mentioned without causing undue distress.

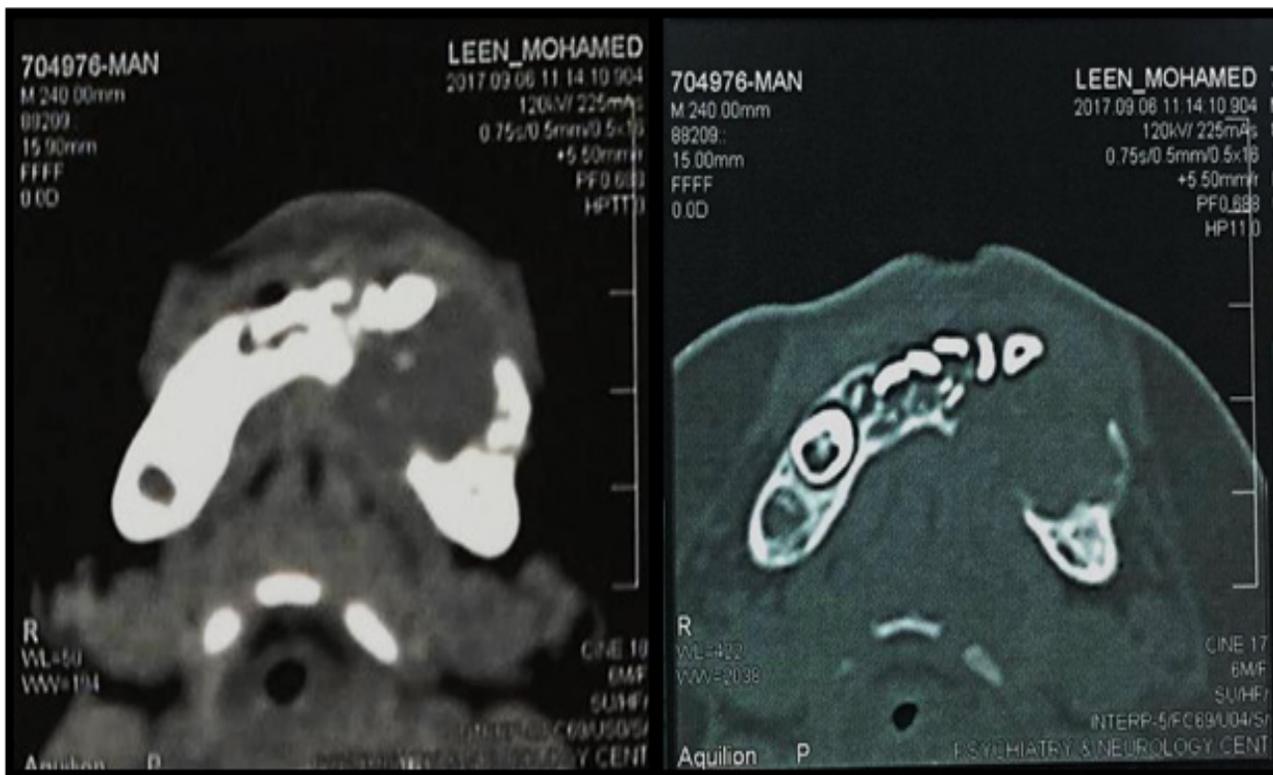
Extra-oral examination showed diffuse firm swelling at left body of mandible without any associated palpable lymph nodes. Head and neck examination revealed no other lesions, or any abnormalities and the general physical examination was within normal limits.

Intra-orally, a painless expansible lesion at the left body of mandible was detected. The lesion exhibited bicortical expansion of both buccal and lingual cortices with no mucosal surface ulceration (Figure 1).

Radiographically, axial and coronal computerized tomography (CT) showed a well-defined unilocular osteolytic lesion at the left body of mandible extending into angle region, measuring about 3 × 2.5 cm with areas of destruction in buccal and lingual cortices (Figures 2, 3).



**Figure 1:** A 6 month old girl intraoperative view showing expansile swelling at the left mandibular posterior region.



**Figure 2:** Axial CT demonstrating a well-defined unilocular radiolucent lesion expanding and perforating buccal and lingual cortices.

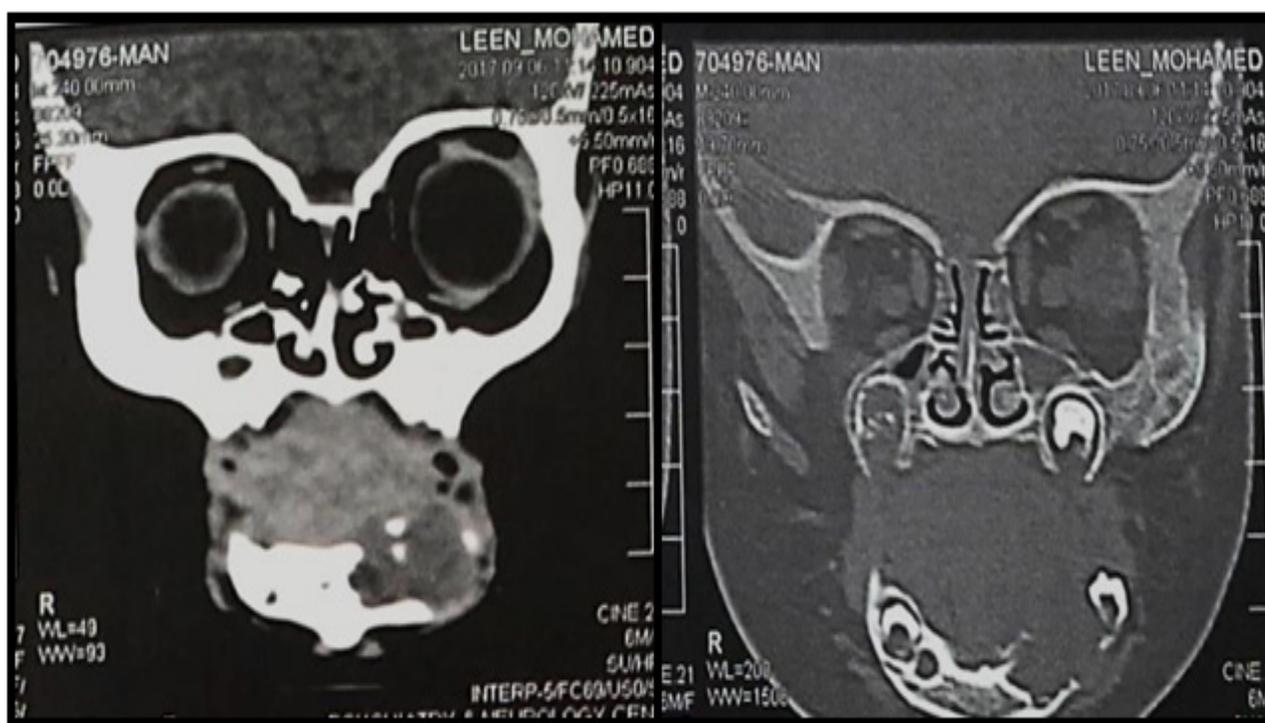


Figure 3: Coronal CT illustrating the broad expansion of buccal and lingual cortices with areas of destruction.

### Surgical procedures

Incisional biopsy was performed under local anesthesia which established the diagnosis of myofibroma. Accordingly, conservative surgical excision of the lesion was planned based upon patient's age, location and size of the lesion.

Under general anesthesia, nasotracheal intubation was done. Following infiltration local anesthesia, buccal and

lingual mucoperiosteal flaps were reflected extending from anterior mandible to anterior border of mandibular ramus and the lesion was exposed.

Complete surgical excision along with peripheral osteotomy was performed with thorough bone curettage before primary closure of the mucoperiosteal flaps. The excised mass was sent for histopathologic analysis (Figure 4).

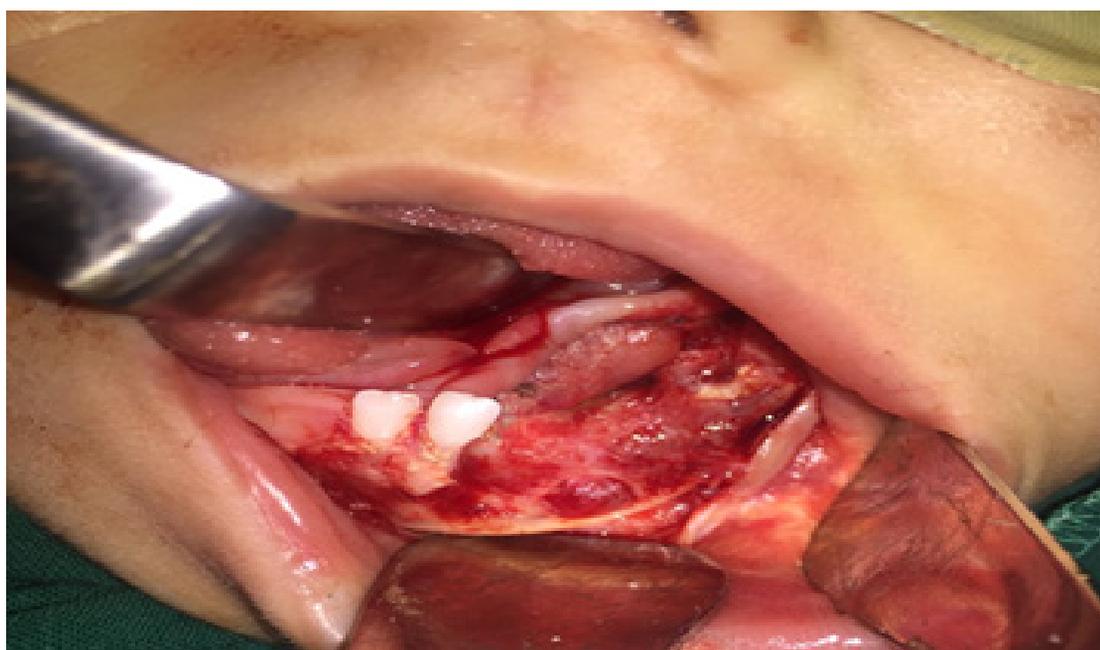
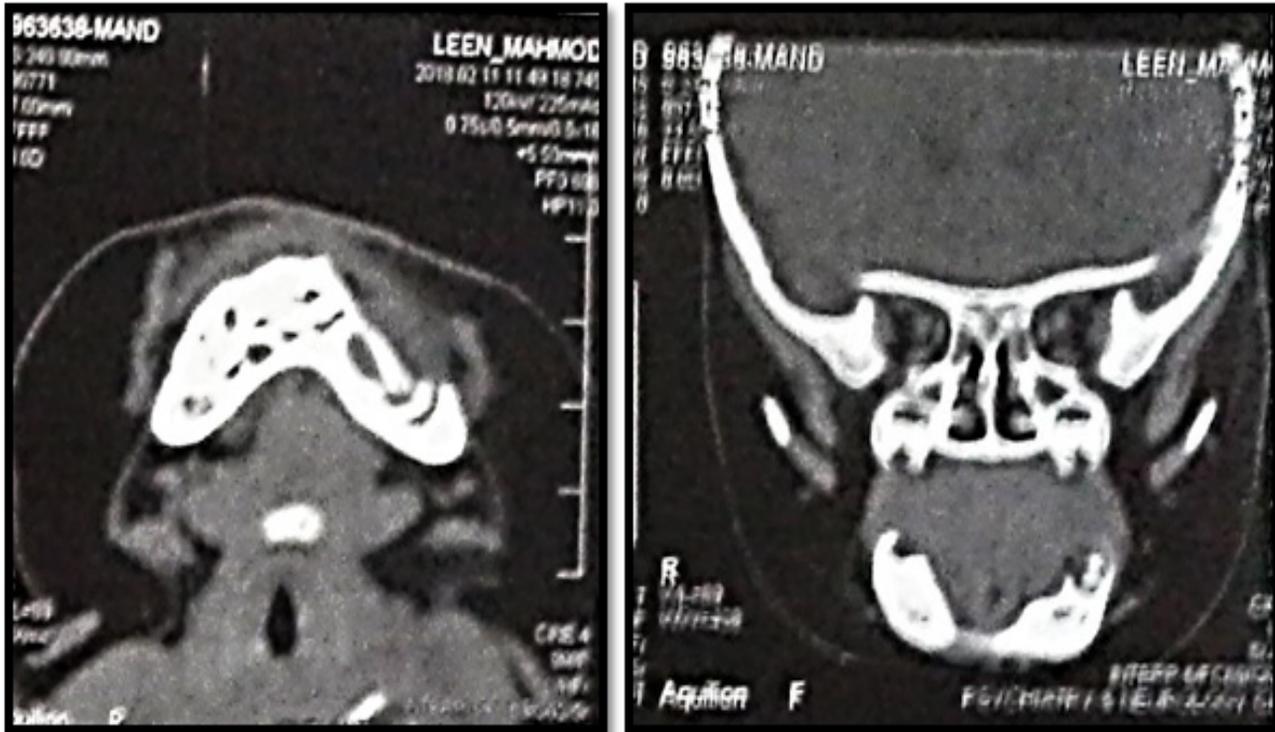


Figure 4: Intraoperative view after complete surgical excision along with peripheral osteotomy of the left posterior mandible.

The patient's postoperative course was uncomplicated and had an uneventful wound healing and 24 months

follow-up period showed no signs of recurrence (Figure 5).



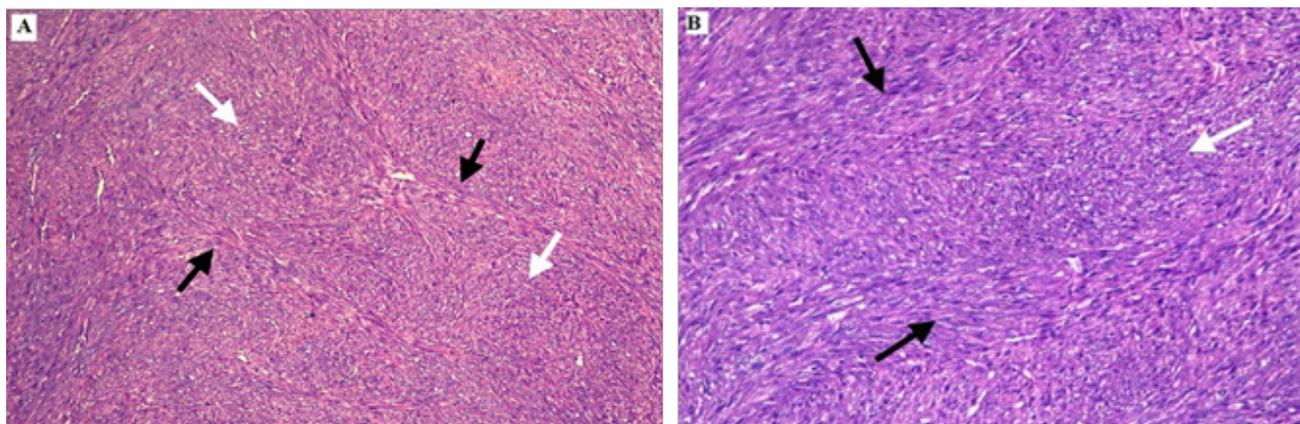
**Figure 5:** Postoperative axial and coronal CT showing an uneventful bone healing with no signs of lesion recurrence.

### **Microscopic findings**

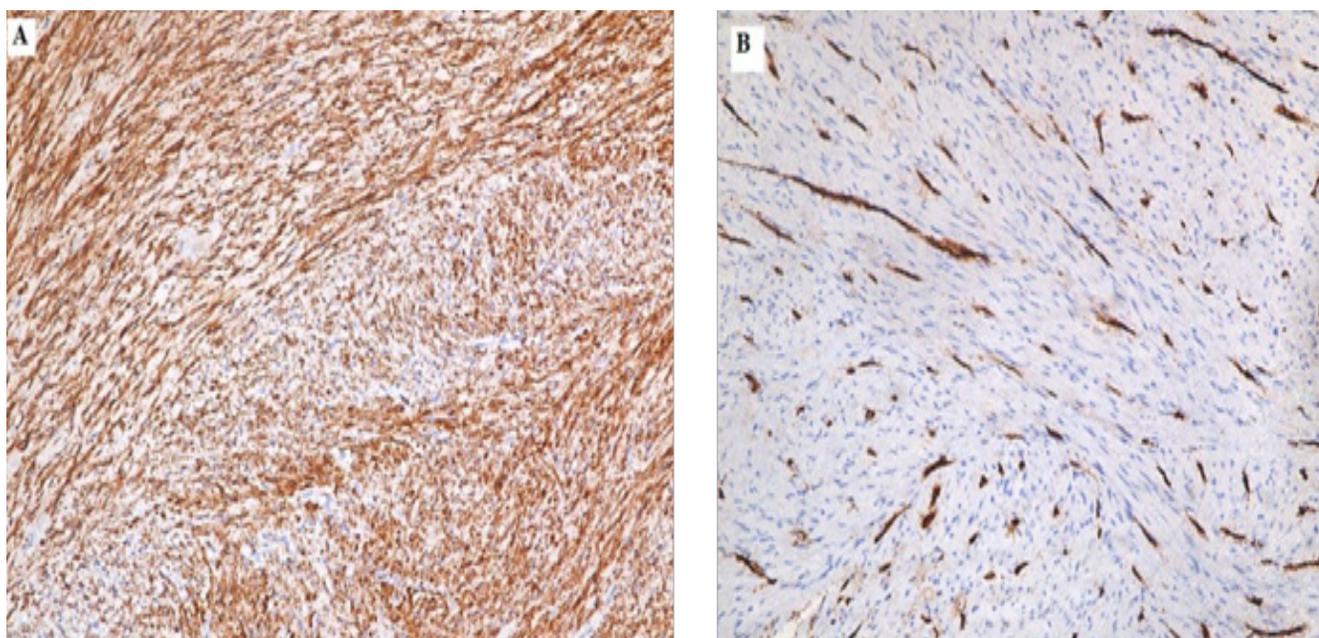
Histopathological examination revealed highly cellular, un-encapsulated mass of spindle-shaped cells with vesicular nuclei and lightly eosinophilic cytoplasm arranged in long, intertwining fascicles. In between the streaming fascicles are densely-packed aggregates of smaller, round to polygonal cells with more compact, basophilic nuclei and little cytoplasm imparting a biphasic or zonal architecture (Figure 6 (A and B)). Some cells exhibited perinuclear, cytoplasmic vacuoles. Numerous slit-like vascular channels were noticed traversing the cellular mass.

The cellular features together with the alternating zones of spindle and round cells were highly suggestive of myofibroma.

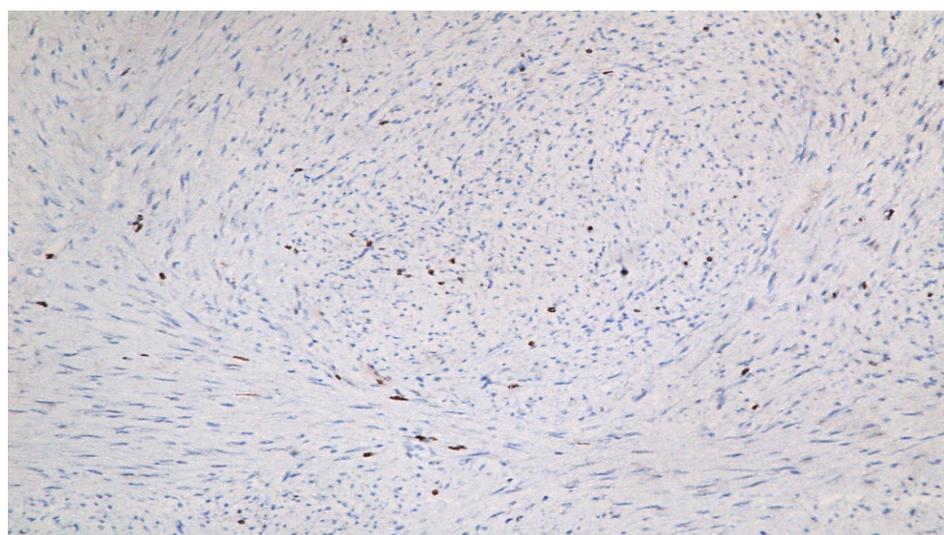
For confirming the diagnosis, further serial sections were prepared for immunohistochemical studies. Immunohistochemistry revealed strong cytoplasmic staining for  $\alpha$ -smooth muscle actin ( $\alpha$ -SMA) (Figure 7.(A)) and negative staining for desmin and S-100 protein. CD34 antibody yielded positive reaction only in endothelial cells rimming the numerous vascular channels (Figure 7 (B)). Tumor cells showed low proliferative index as estimated by the few positive Ki67-labelled cells (Figure 8).



**Figure 6:** (A) Streaming fascicles of spindle-shaped cells (black arrows) admixed with foci of densely-packed rounded cells (white arrows). (B) Higher magnification clarifying the zoning phenomenon with blended lighter-stained (black arrows) and darker-stained (white arrow) areas (H&E; original magnification (A)  $\times 20$ , (B)  $\times 40$ ).



**Figure 7:** (A) Positive immunostaining with  $\alpha$ -SMA. (B) CD34 highlights intralésional blood vessels whereas no reactivity is observed in tumor cells (ABC; original magnification (A) and (B)  $\times 40$ ).



**Figure 8:** Positive nuclear immunostaining for ki67 in sporadic tumor cells (ABC; original magnification  $\times 20$ ).

## DISCUSSION

Myofibroma is a rare benign mesenchymal neoplasm of myofibroblasts that may present clinical and histopathologic features overlapping with several benign and malignant spindle cell tumors. The tumor usually follows an indolent course, afflicting both adults and children most commonly in head and neck region, trunk and extremities but rarely within bone. Intraosseous myofibroma exhibits a wide spectrum of biologic behavior ranging from very mild to moderate degrees of invasiveness<sup>[1]</sup>.

The present case illustrates a central myofibroma in a 6-month-old girl. This is in line with the notion that oral myofibromas mostly predominate among females

during the first two decades of life. In 2017, Smith *et al.*<sup>[1]</sup>. Performed a literature review of more than 200 cases where they reported that most of intra-osseous myofibromas occurred in patients under the 20 years of age while the majority of patients over 40 years of age had extra-osseous, mucosal lesions<sup>1</sup>. Radiographically, central myofibroma occurring in pediatric patients usually present as a unilocular radiolucency sometimes involving the cortical or periosteal surfaces<sup>[1, 6]</sup>. The radiographic appearance of our case was conforming to this description. Since its similarity to several benign and malignant lesions, careful microscopic and immunohistochemical examination of biopsied tissue is of utmost importance for establishing the definitive diagnosis.

The histopathologic features of our case were in accordance with those described for oral myofibromas<sup>[1, 6]</sup>.

To further exclude its potential histopathologic mimics such as leiomyoma, schwannoma, solitary fibrous tumor and infantile fibrosarcoma, immunohistochemical studies were carried out. Our case revealed negative staining with desmin and S-100 protein which precluded the possibilities of leiomyoma and schwannoma respectively. Negative staining with CD34, except for lesion vessels, excluded the possibility of solitary fibrous tumor.

The positive cytoplasmic reaction for  $\alpha$ -SMA confirms the myofibroblastic nature of tumor cells and the low Ki67-labelling index further augment the benign nature of this lesion. Matthews et al. have advocated that intraosseous jaw lesions should be treated by thorough curettage or further wide excision where extension into soft tissue is identified<sup>[12]</sup>.

We intended to preserve the lower border and remnants of both buccal and lingual cortices as the lesion was benign and the preoperative CT showed a residual sound bone and to avoid plating that might interfere with growth and need to be removed in another general anesthesia set.

## CONCLUSION

Despite being rare in the mandible, infantile solitary myofibroma can sometimes represent a diagnostic dilemma since its radiographic similarity to a number of aggressive lesions. Hence, definitive preoperative diagnosis is of substantial clinical importance as the lesion responds favorably to simple conservative excision and the patient remains disease free at 24 months follow-up period.

## CONFLICT OF INTEREST

There are no conflicts of interest.

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