

# Myofibroma of the oral cavity mimicking a non-neoplastic inflammatory process: an unusual report

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## Abstract:

Myofibroma is a rare condition that is a benign neoplasm characterized by the proliferation of fusiform cells originated from myofibroblasts. A 9-year-old male patient presented a intraosseous lesion in the oral cavity, which caused mobility in two teeth within approximately 1 month and was submitted to excisional biopsy. Microscopic analysis revealed fragments of neoplasia of mesenchymal origin, characterized by proliferation of spindle cells with eosinophilic staining, showing loose chromatin and indistinct cytoplasmic boundaries, as well as cells with vacuolated cytoplasm. The lesion was very cellular and organized into a bundle of intersecting neoplastic cells. In the immunohistochemical analysis, the proliferative index evaluated for the Ki-67 antibody was below 5%. Neoplastic cells were strongly positive for the smooth muscle actin antibody. Rare mitoses were observed in the specimen, leading to the diagnosis of myofibroma. After the diagnosis, the patient was referred to his pediatrician for investigation of possible myofibromatosis. After clinical and imaging analysis, no similar lesions were found, and the diagnosis of myofibromatosis was excluded. At 6 months postoperative, the patient returned to the outpatient clinic, without clinical and radiographic signs of recurrence and buccal tissues with normal appearance.

**Keywords:** myofibroma; oral cavity; oral pathology; immunohistochemistry

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## INTRODUCTION

Myofibroma was first described by Stout in 1954 as “congenital generalized fibromatosis”<sup>1</sup>. It is a benign neoplasm characterized by the proliferation of myofibroblasts, which are fusiform<sup>1,2</sup>, commonly observed in children: 89% of cases occur before 2 years of age, and, of these, 54% are already present at birth. The most affected sites are skin, bones, lungs and gastrointestinal tract<sup>1</sup>.

The appearance of solitary lesions is more common, and when it affects multiple sites this condition is called myofibromatosis, characterized by lesions found in the lungs, kidneys, pancreas, gastrointestinal tract and bones<sup>1,3</sup>. The exact etiology of this condition is unknown, most cases are reported as sporadic, although some studies suggest the possibility of a familial pattern, suggesting that myofibromatosis may be inherent in an autosomal dominant or recessive trait<sup>4,5</sup>. The presentation in the disseminated form has a poor prognosis, so an adequate clinical examination is important to rule out the presence of myofibromatosis.

In general, myofibroma is asymptomatic and the most reported clinical presentation is the swelling, and, to a lesser degree, the presence of an exophytic oral lesion involving the alveolar ridge and gingiva<sup>3,6,7</sup>. Therefore, displacement and tooth mobility may occur. When these lesions affect the mandible, it has the potential to affect teeth and other odontogenic structures. In these cases, clinical and radiographic findings may suggest an odontogenic cyst or tumor, such as ameloblastoma, keratocyst, dentigerous cyst, benign and malignant central neoplasias and odontogenic fibromyxomas<sup>4</sup>, hence diagnosis can be challenging.

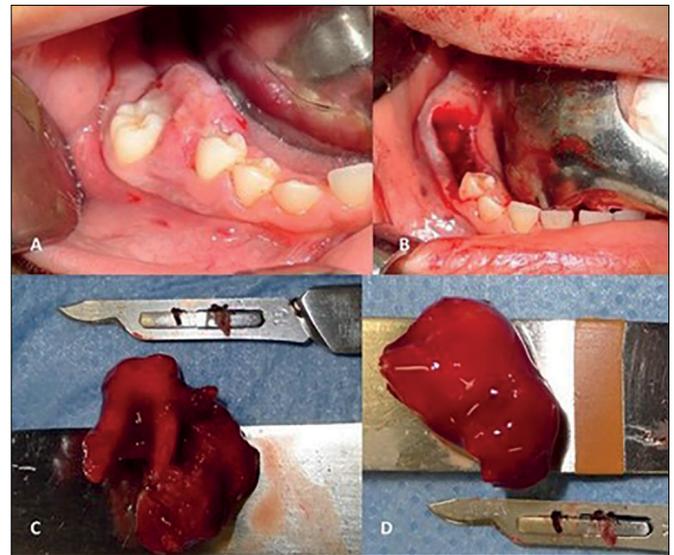
Due to the low incidence of myofibromas in the oral cavity and its rare nature, the purpose of this article is to report a case of this lesion in the posterior region of the mandible, focusing on the histopathological diagnosis.

## CASE REPORT

A 9-year-old male patient was brought by his parents to the Oral and Maxillofacial Surgery clinic at the Municipal Hospital “Dr. Mário Gatti” in Campinas - SP, after they had observed the presence of a lesion in the oral cavity, which caused mobility in two teeth within approximately 1 month. The patient was healthy and there were no significant findings in prior medical history, including medication use, allergies, and family

history. The lesion showed a rapid growth and, according to his relatives, there was no causative etiological factor, such as a trauma or an infection.

During the clinical examination, an ill-defined, sessile nodal mass was observed. It had a smooth and shiny surface in the region of the right mandibular alveolar ridge, and caused lateral and coronal displacement of the lower right 1st molar and lower right 2nd premolar resulting in slight extrusion (Figure 1A). Upon palpation, the lesion had a fibroelastic consistency which was painless and non-bleeding, in addition to marked mobility in the latero-lateral directions of the aforementioned teeth. The patient had satisfactory oral hygiene and there were no signs of cervical lymphadenopathy.

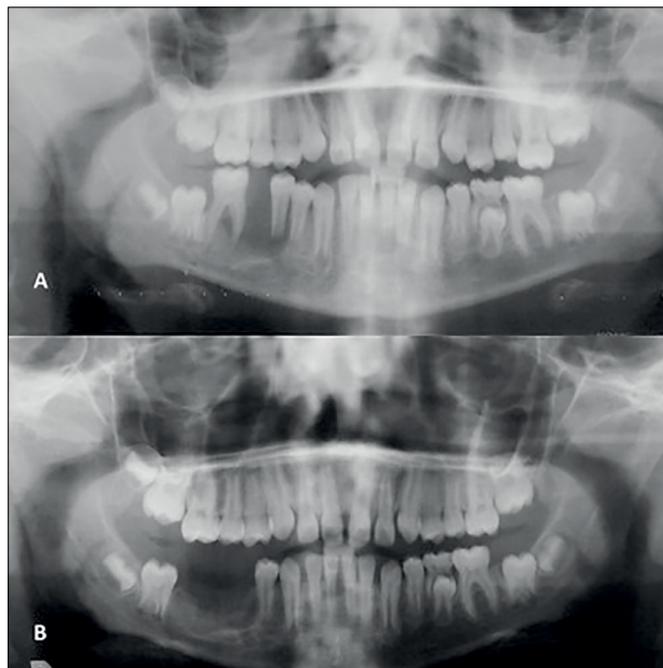


**Figure 1.** A- Preoperative appearance of nodular lesion, with ill-defined borders, sessile, discretely reddish, smooth and shiny surface in the region of the right mandibular alveolar. B- Surgical cavity after cleavage and lesion excision. C & D - Lesional aspect after resection.

Through radiographic examination, a non-corticalised radiolucent unilocular lesion was observed, with well-defined limits, leading to a significant reabsorption of the alveolar bone around the permanent lower right 1st molar and lower right 2nd premolar, as well as extrusion thereof (Figure 2A). Based on the clinical and radiographic data, a provisional diagnosis of peripheral lesion of giant cells or some more aggressive form of pyogenic granuloma were suggested.

The excision of the lesion was performed under general anesthesia, for better patient comfort. The procedure started with anesthetic infiltration and aspiration to certify that the lesion had no vascular origin. The lesion was easily removed after incision

with an electric scalpel and dissection with periosteal elevators (Figure 1B). The involved teeth were also removed (Figure 1C, D). Thereafter, bone curettage was performed to eliminate any tumor remnants. After copious irrigation with saline solution, the primary closure of the wound was performed with resorbable sutures.

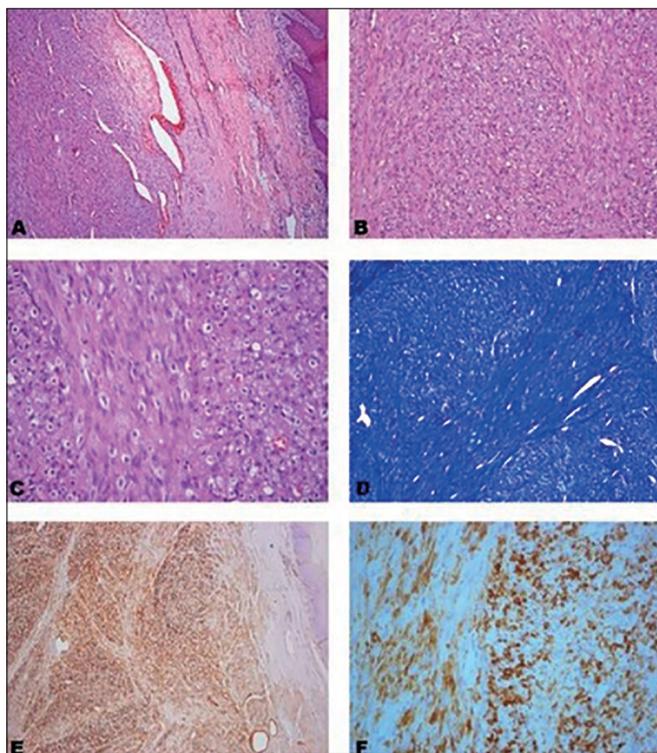


**Figure 2.** A - Radiographic aspect of unilocular lesion, with well-defined limits, leading to a significant reabsorption of the alveolar bone around the permanent lower right 1st molar and lower right 2nd premolar. B - Postoperative radiograph showing absence of recurrence.

In the outpatient returns, the patient did not present cicatricial alterations, infections or any other postoperative complication. The surgical specimen was sent to the Oral Pathology Laboratory of São Leopoldo Mandic College, Campinas - SP.

In the macroscopic analysis, 02 fragments of soft tissue were observed. The largest fragment measured 24mm x 13mm x 10mm and the smallest one 22mm x 15mm x 09mm, with irregular shape and surface, pink and blackish coloring and fibrous consistency. Microscopic analysis revealed fragments of mesenchymal origin, characterized by zoning phenomenon that it exhibited peripheral spindle cells with oval to tapering nuclei, whereas the central round to polygonal cells had scant cytoplasm and hyperchromatic nuclei. Abnormal mitotic figures and areas of necrosis were absent. It was possible

to observe a stroma composed of dense connective tissue and small blood vessels (Figure 3A, B, C). A special stain technique for collagen using Masson's trichrome stain was used to quantify the stromal component within the lesional mass. The special stain demonstrated an excess of collagen (blue) around the cellular component (pink) (Figure 3D). In the immunohistochemical analysis, the proliferative index evaluated for the Ki-67 antibody was below 5%. Neoplastic cells were strongly positive for the smooth muscle actin antibody (Figure 3E, F). Rare mitoses were observed in the specimen, leading to the diagnosis of myofibroma.



**Figure 3.** (A, B & C) Hematoxylin and eosin stained section shows fascicular and cellular areas characterized by polygonal cells at the center and elongated cells at the periphery. (D) Masson's trichrome stain highlights the highly fibrous stroma. (E & F) Immunohistochemical stain tumor cells are positive for smooth muscle actin marker.

After the diagnosis, the patient was referred to his pediatrician for investigation of possible myofibromatosis. After clinical and imaging analysis, no similar lesions were found, and the diagnosis of myofibromatosis was excluded. At 6 months postoperative, the patient returned to the outpatient clinic, without clinical and radiographic signs of recurrence and buccal tissues with normal appearance (Figure 2B).

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## DISCUSSION

Myofibroma and myofibromatosis are rare spindle cell neoplasms, composed of myofibroblasts<sup>5,8</sup>. These cells have intermediate characteristics between fibroblasts, undifferentiated cells and smooth muscle. These lesions commonly appear in children and infants (average 7.2 years old)<sup>2,5</sup>, although they may develop at any age<sup>3,4,5,8</sup>.

The cause of myofibroma is actually unknown, although trauma has been implicated in the pathogenesis of this lesion. In response to trauma or any mechanical tension the fibroblast cells get stimulated and under the influence of platelet derived growth factor they are differentiated into protomyofibroblast. In the cytoplasm of this cell, accumulation of more amounts of actin microfilaments occurs. Then under the influence of transforming growth factor- beta and EDA-Fibronectin these protomyofibroblast cells are differentiated into myofibroblast cells<sup>9</sup>.

Clinically, these lesions tend to appear the head and neck, with oral lesions affecting mainly the mandible, lips, cheek and tongue<sup>5</sup>. Among the 79 cases of myofibroma of the oral cavity analyzed by Foss and Ellis<sup>10</sup>, it was reported that the most common site was the mandible (n = 30), followed by the lips (n = 14) buccal mucosa (n = 14), tongue (n = 13), maxilla (n = 4), pterygomaxillary space (n = 2), oral floor (n = 1) and submandibular gland (n = 1). The case reported in this paper presents clinical data in agreement with the literature findings, once the patient was diagnosed in the first decade of life and the lesion developed in the mandible.

Although this lesion is a benign neoplasm, a varied biological behavior has been observed, ranging from mild to moderate infiltrative capacity<sup>1,2,8</sup>. The patient of this report had an exophytic lesion, fast-growing, causing mobility of the associated teeth. These clinical findings are compatible with myofibroma, in which is possible to observe painless swelling that can result in cortical perforation, sometimes presenting secondary ulceration and root resorption<sup>5,8,10,11</sup>. These aspects may induce the suspicion of malignant neoplasms or other locally aggressive lesions. The absence of symptoms, even when the lesions expand and / or puncture the cortical and manifest as an oral mass, is an important factor in the delay of the diagnosis<sup>11,12</sup>.

Myofibromatosis and myofibroma are histopathologically identical. However, there may be a predilection for localization. Myofibromatosis can occur in the dermis, subcutaneous tissue, muscle, skeleton and internal

organs, while solitary myofibromas are more common in the dermis and subcutaneous tissue<sup>13</sup>. Myofibromatosis involving multiple organs has a poor prognosis due to severe cardiorespiratory and gastrointestinal complications, which often make it difficult to manage patients and lead to poor overall health status<sup>2</sup>. Therefore, it is important to perform anamnesis and clinical examinations to exclude the presence of multiple lesions. After being diagnosed with myofibroma, the patient was referred to his pediatrician, who discarded the hypothesis of myofibromatosis.

Previous reports have described myofibroma as a radiolucent unilocular lesion on radiographic examination, with well-defined borders, findings similar to those found in the radiographic examination of the patient of this report<sup>3,4,5,8,10,11</sup>. Although lesions can be observed in the multilocular aspect, with cortical expansion and/or perforation<sup>3,5,8</sup>. In the literature review performed by Allon et al.<sup>11</sup> of the 16 cases in which radiographic information was possible,<sup>12</sup> (75%) presented involvement of the tooth or dental germ. Computed tomography can also be used as a tool to diagnose these lesions<sup>3</sup>. In this report, the use of tomography was not necessary, since the panoramic radiograph provided sufficient information regarding the characteristics of the lesion, its limits and involvement of adjacent structures.

The most variable aspect of this tumor is the histopathological aspect. Microscopically, the lesion presents as a well-circumscribed spindle cell neoplasm which exhibits a typical zonal appearance characterized by elongated peripheral cells arranged in short fascicles or spirals and a center surrounded by polygonal-shaped cells<sup>5,8,10,11,13,14</sup>. The biphasic nature of this lesion is caused by the longitudinal and cross-section of the spindle cells, which can also result in a characteristic organoid pattern<sup>4,5,8</sup>. The histopathological differential diagnosis includes a spectrum of non-odontogenic neoplasms of mesenchymal origin such as: leiomyoma, fibrosarcoma, benign nodular histiocytoma, solitary fibrous tumor and hemangiopericytoma<sup>1,2,3</sup>. The immunohistochemistry technique, together with the morphological aspect, is very useful in distinguishing between the different lesions that make differential diagnosis with the myofibromas. The neoplastic cells of myofibromas show strong and diffuse expression pattern for smooth muscle, muscle specific actin and vimentin antibodies and negativity, with rare exceptions, for S100, desmin, CD34, CD99 and cytokeratins<sup>2,3,8,14,15</sup>.

Two lesions that may exhibit very similar morphological and immunohistochemical characteristics are the solitary fibrous tumor and nodular fasciitis. In the case of solitary fibrous tumor, the positivity observed in the neoplastic cells for CD34, CD99 and Bcl-2 allows the establishment of the diagnosis 15. In the case of nodular fasciitis, because it is an lesion consisting of myofibroblastic and fibroblastic cellular components, the immunohistochemical profile may be very similar to that observed in myofibromas, with positivity for smooth muscle actin, muscle specific actin and vimentin. However, growth pattern and morphological characteristics are useful in distinguishing between the two lesions. The distinction between sarcomas that may affect the orofacial region should be based on the absence of pleomorphism, atypia, atypical mitoses and high mitotic index in cases of myofibromas. In the present case, a proliferative index below 5% and absence of the histological parameters mentioned previously were observed, which allowed to avoid the possibility of malignant lesion.

Despite being a well-recognized entity, myofibromas are often misdiagnosed<sup>4,11, 3</sup> because of their infiltrative nature. Undiagnosed cases may contribute to a decrease in incidence and lead to a mistaken treatment. Wrong diagnoses generally include spindle cell lesions of the nervous tissue or originated from the smooth muscle, and more aggressive myofibroblastic lesions, desmoplastic fibroma and low grade fibrosarcoma<sup>11</sup>. In particular, leiomyosarcoma may show a similar infiltrative pattern and may have few mitoses, but will generally show a more uniform fusiform morphology, will exhibit cytologic atypia, and will be positive for desmin<sup>4</sup>. Foss and Ellis 10 reported that 7 of 16 cases of myofibromas were initially diagnosed as other diseases.

The most common modality of treatment is local excision<sup>2,5,8,14</sup>. Corroborating with the series of cases reported by Smith et al.<sup>15</sup>, in which conservative treatment was the most used, the proposed treatment was effective, since the patient had no evidence of recurrence in the sixth postoperative month. The prognosis varies according to the site of tumor occurrence, the recurrence rate varies from 7 to 31% and is more likely in cases where surgical access is more difficult<sup>8</sup>.

## CONCLUSION

Myofibroma is a neoplasm that can affect children and adolescents, with a predilection of the

mandible when the head and neck are involved. Thorough clinical examination is essential to rule out myofibromatosis. Anatomopathological analysis and immunohistochemical evaluation were essential to rule out malignancy of the lesion. This is important for the administration of adequate treatment that is, an appropriate resection with accurate tumor free margins.

**Compliance with Ethical Standards Conflict of interest:** The authors declare that they have no conflict of interest.

**Ethical Approval:** For this type of study formal consent is not required.

**Informed Consent:** Informed consent was obtained from all individual participants included in the study.

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