# Infantile myofibroma of the mouth floor

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## ABSTRACT

**AIM:** Infantile myofibroma is a rare benign solitary tumor of mesenchymal origin which rarely occurs in the mouth floor. Our aim was to emphasize the importance of the histological and immunohistochemical features and the potential diagnostic challenges of infantile myofibroma.

**CASE REPORT:** A 7-year-old boy presented to consultation for a swelling of the mouth floor. It was 4 cm in diameter, solid, painless and well limited tumor. Maxillofacial computed tomography showed an homogenous tissular mass. A complete excision was performed. The histology and the immunochemistry concluded to the diagnosis of a myofibroma. Evolution was uneventful and after 24 months of follow-up, there was no relapse.

**CONCLUSION:** Infantile myofibroma might be clinically alarming, because of the rapid growth or the location. For precise diagnosis, both histopathological examination and immunochemistry are necessary. The treatment is total tumor excision.

**KEYWORDS:** Infantile Myofibroma, Mouth Floor, Immunochemistry, Surgery, Prognosis.

## INTRODUCTION

Myofibromatosis is the most frequent fibrous tumor in childhood, A myofibroma is defined as a solitary tumor which is a rare mesenchymal disorder that occurs usually in the subcutaneous, cutaneous or muscular tissues [1]. The report of this particular case of a solitary lesion involving the mouth floor aims to emphasize the importance of the histological and immunohistochemical features and the potential diagnostic challenges.

## CASE REPORT

A 7-year-old-boy, with no significant medical history, presented with a one year history of a slow growing mass of the mouth floor. This swelling was painless and did not affect his speech. Physical examination revealed a mass located in the anterior part of the mouth floor measuring 4 cm in diameter (Figure 1). It was hard with palpation, irregular, well-circumscribed and lifting the tip of the tongue with a prolongation downward. There was not cervical lymphadenopathy.

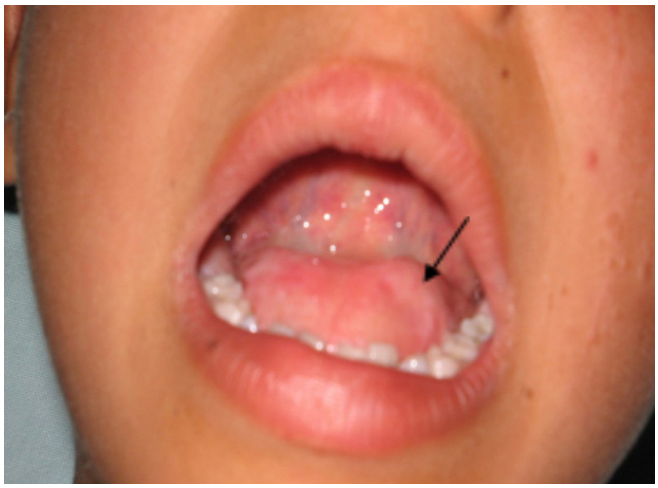

**Figure 1 :** Preoperative view of the mouth floor

Facial computed tomography scan showed a well-defined homogeneous mass of the anterior floor, of a soft tissue density with an intra-tumoral calcification. There were no bone erosion or contrast enhancement (Figure 2).

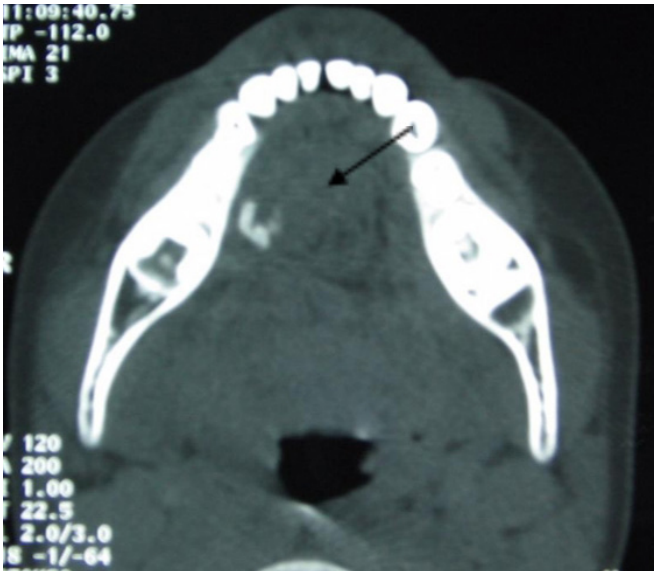


**Figure 2 :** Computed tomography scan: Tumor with soft tissue density associated to a lateral calcification

The preliminary diagnosis was dolichomega-salivary submandibular duct or benign fibrous tumor. Complete surgical resection was easily performed under general anesthesia. Gross specimen of the the well defined fibrous tumor was 5 cm x 4 cm (Figure 3).

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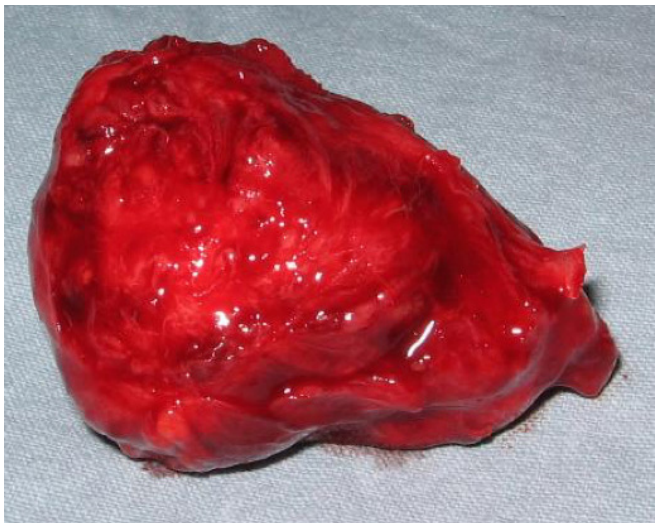

**Figure 3 :** Macroscopic aspect of the specimen

The histological examination concluded that it was a myofibroma (Figures 4).

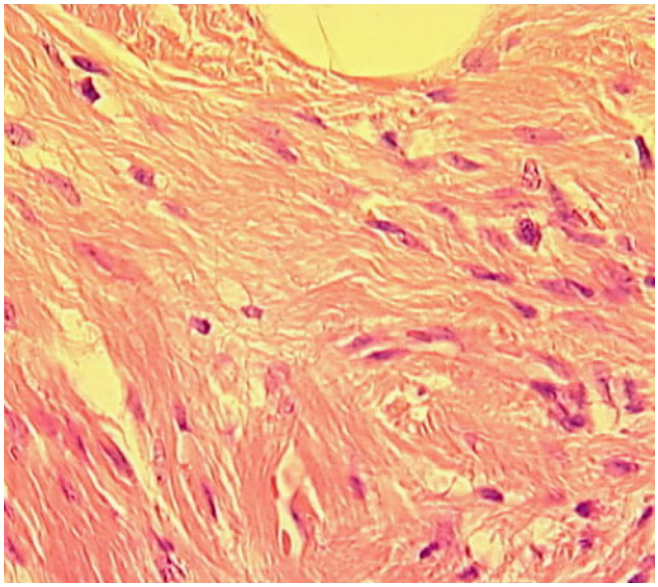

**Figure 4 :** Histopathological examination showing regular myofibroblasts (haematoxylin and eosin stain, 400 x magnification).

Immunochemistry was positive for vimentin. The follow-up showed no complications and there was no sign of recurrence after two years.

## DISCUSSION

Fibromatosis represent a group of infiltrating fibrous proliferations of fibroblasts, myofibroblasts, or both that exhibit a clinical behavior and biologic potential intermediate between benign and malignant fibrous lesions [2]. Although rare, the myofibromatosis are the most frequent type of these tumors in infancy [3].

They were first described in 1954 by Stout as, congenital generalized fibromatosis. Then, in 1981 Chung and Enzinger defined the histopathological features of the disease in infants and used the term, infantile myofibromatosis [1]. Most cases present during the first year of life [4] with a slight male predominance [3].

The most common site of involvement for these lesions in children is the head and neck region where the lesions affect most frequently the dermis, subcutaneous tissues, and muscular structures [5]. The involvement of the mouth floor, which may lead to obstructive complications, is rarely described [2]. Clinically, three different varieties of infantile myofibromatosis are described with different prognosis; solitary, multicentric, and multicentric with visceral involvement. The solitary form is now recognized as the most common presentation (74%) [3]. The term myofibroma is referred to this form and typically, presents a benign clinical evolution with a slow growing mass [6, 7, 8]. Radiological examination has only supportive role in diagnosis, but do not suggest a definitive diagnosis as infantile myofibromatosis does not have a characteristic appearance. However, imaging can be helpful in evaluating the extent of the disease, the progression, the regression and the recurrence. The computed tomography shows an homogeneous and well-contrasted tumor. Necrosis and intratumoral calcification might be seen [3, 6]. Precise diagnosis is, generally, made after histopathological examination. It shows spindle-shaped cells, arranged in bundles or fascicles and that appear intermediate between fibroblasts and smooth muscle cells. At its center, perivascular round cells are usually observed [5]. However, the microscopic appearance of these tumors often does not reflect the benign clinical characteristics. Thus, this entity needs to be differentiated from other tumors of infancy, including neurofibromas, agressive fibromatosis, infantile fibrosarcoma, fibrous histocytoma and leiomyosarcoma. Histological features consistent with malignancy such as local invasion, increased cellularity, rich vascularisation, and extensive necrosis, are not uncommon [1, 9]. Neither metastasis nor malignant degeneration of myofibromas, however, has been reported [1]. Besides, immunochemistry is useful to differentiate this tumor from other spindle cell fibroblastic tumors especially the aggressive fibromatosis that has a higher rate of recurrence and may degenerate in a fibrosarcoma [5, 9]. It shows a positivity of the myofibroblastic component for vimentine and alpha-smooth muscle actin, while both desmin and S-100 protein are negative [8]. Familial clustering of infantile myofibromatosis cases has suggested genetic heritability transmitted in both autosomal dominant or autosomal recessive patterns with variable penetrance, but, no concrete genetic basis has been determined yet [4].

For the solitary type of infantile myofibromatosis, complete excision of the tumor is curative [2, 5]. The rate of recurrence is between 7 and 31% [10].

## CONCLUSION

Myofibromas represent the most frequent type of fibrous neoplasm in infants. They commonly occur in the head and neck region but, the mouth floor is a rare location. Therefore proper correlation of clinical and findings with histologic morphology and immunochemistry is necessary for

diagnosis. The primary treatment is surgical and long-term follow-up of the patient is mandatory due to the possibilty of recurrence.

**Competing interests:** The authors declare no conflicts of interest.

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