Childhood Intraosseous Myofibroma: A Case Report and Review of the Literature

Çocukluk Çağı İntraosseöz Myofibroma: Bir Olgu Sunumu ve Literatürün Gözden Geçirilmesi

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ABSTRACT

Myofibroma, a rare benign tumor, frequently occurs in the head and neck regions, particularly the tongue, gingivae, maxillary, and mandible, often involving the bone. This tumor comprises highly proliferative spindle cells, including fibroblasts and myoblasts, and often presents with lytic radiographic areas and painless masses. Treatment varies from conservative management to radical resection, depending on patient age, tumor location, and severity. This report details a rare case of mandibular myofibroma with bony involvement in a 6-year-old girl, highlighting its diagnosis and treatment. The patient presented with a painless, smooth red mass in the right lower jaw, impairing chewing and causing bleeding. Clinical and radiological evaluations revealed a fixed mass between the right primary canine and permanent first molar, with radiolucent features on panoramic radiography. An excisional biopsy under general anesthesia was performed, leading to the removal of the lesion and the first molar. Histopathological examination identified spindle cells with myofibroblastic differentiation, and immunohistochemical staining showed a high proliferative rate of Ki67. Follow-up over two years showed no recurrence, and soft tissue healing was satisfactory. This case emphasizes the importance of conservative management and thorough follow-up in treating pediatric myofibroma with bony involvement, avoiding radical procedures where possible.

Keywords: *Myofibroma, Oral Pathology, Mandibular Neoplasms*

ÖZ

Nadir görülen benign bir tümör olan myofibroma sıklıkla baş ve boyun bölgelerinde, özellikle dil, gingiva, maksilla ve mandibulada görülür. Mandibular kemik tutulumu olan myofibroma çocuklarda nadir görülmektedir. Bu tümör, fibroblastlar ve myoblastlar dahil olmak üzere yüksek oranda proliferatif iğsi hücrelerden oluşur ve genellikle litik radyografik alanlar ve ağrısız kitlelerle kendini gösterir. Tedavi hastanın yaşı, tümörün yeri ve şiddetine bağlı olarak konservatif tedaviden radikal rezeksiyona kadar değişir. Bu raporda, 6 yaşında bir kız çocuğunda kemik tutulumu ile seyreden nadir bir mandibular myofibroma olgusunun tanı ve tedavisi anlatılmaktadır. Hastamızda sağ alt çenede ağrısız, düzgün yüzeyli, kırmızı renkli, çiğnemeyi bozan ve kanamaya neden olan bir kitle saptandı. Klinik ve radyolojik değerlendirmelerde sağ süt kanin ile daimi birinci büyük azı dişi arasında panoramik radyografide radyolusent özellikler gösteren bir kitle tespit edildi. Genel anestezi altında eksizyonel biyopsi yapılarak lezyon ve birinci molar diş çıkarıldı. Histopatolojik incelemede myofibroblastik farklılaşma gösteren iğsi hücreler tespit edildi ve immünohistokimyasal boyamada yüksek Ki67 proliferatif oranı görüldü. İki yıl boyunca yapılan takiplerde nüks görülmedi ve yumuşak doku iyileşmesi iyi sonuç verdi. Bu vaka, kemik tutulumu olan pediatrik myofibroma tedavisinde mümkün olduğunca radikal prosedürlerden kaçınarak konservatif yönetimin ve kapsamlı takibin önemini vurgulamaktadır.

Anahtar Kelimeler: *Myofibroma, Oral Patoloji, Mandibular Neoplazmlar*

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INTRODUCTION

Myofibroma is a rare condition that is classified according to the body region where it is diagnosed[1-3]. Benign tumour myofibroma is often diagnosed in the head and neck region, followed by the tongue, gingivae, maxillary and mandible[1-4]. This benign tumour, which is defined as multicentric myofibromatosis, has highly proliferative spindle cells, including fibroblasts and myoblasts[5-7].

This tumour has similar appearances among many clinical situations, such as lytic areas on radiography and gingival changes, including a painless mass, when cortical plate perforation occurs[8]. Thus, determining a differential diagnosis is rather difficult[5].

The histological characteristics of Myofibroma are defined by a polylobulated spindle cell proliferation arranged in a biphasic pattern. Immunohistochemical staining reveals that the tumour cells express alpha-smooth muscle actin (α-SMA), while they are typically negative for myogenin, desmin, CD34, S-100 protein, and beta-catenin[9]. However, the immunohistochemical analysis of Ki-67-positive cells in a range of tumour types has been demonstrated to be an effective method for evaluating tumour growth potential[10].

Treatment options are quite extensive, such as conservative management, curettage or radically resective surgery[1-6,11]. The age of patients, localization and involvement of the tumour, and life-threatening conditions are the determining factors for its management[12].

This report describes the diagnosis and treatment of a rare case of myofibroma in a 6-year-old girl on the posterior mandibular region. Attention has been focused on similar histopathological features of this tumour with many oral pathologies and a high proliferative rate (< 15%) of Ki-67 marker, as in this present case, has rarely been reported in literature.

CASE PRESENTATION

A 6-year-old female was referred to Oral and Maxillofacial Surgery Department of the University with a 1-month history of a painless smooth mass with a red colour in the right lower jaw area. The patient's parents had complained of the patient's inability to chew and bleeding of the expanding swelling. The patient had no history of trauma in the recent past, and her routine laboratory tests were normal.

Clinical and radiological evaluation

Clinical evaluation revealed a fixed palpable mass with a height of 2–3 mm from the occlusal level between the right primary canine and permanent first molar and without any associated cervical lymphadenopathy. The clinical appearance of this lesion was a soft tissue lesion resembling a peripheral giant cell granuloma. Panoramic radiography showed a radiolucent lesion with impacted premolars (Figure 1). In addition, cone beam computed tomography was performed to peruse the size and border of this lesion (Figure 2).

Based on the clinical and radiological findings, incisional biopsy under local anaesthesia was planned but due to the patient being uncooperative, the biopsy that was planned for diagnostic purposes could not be performed. Excisional biopsy under general anaesthesia was performed, after the family signed an informed consent agreement.

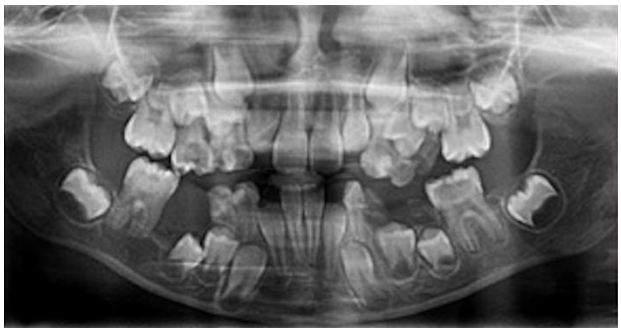

Figure 1: The lenght and size of the lesion on panoramic radiography

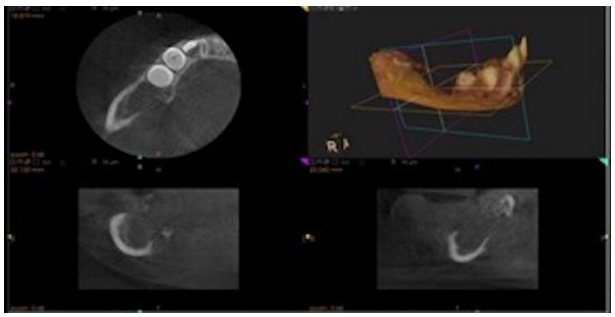

Figure 2: CBCT analysis of the lesion. Lingual plate perforation and extansion of the tumor were observed.

Dental treatment

Primarily, the mucoperiosteal flap was elevated from the mandibular right canine to the first molar. A burr or cutting device was not used, and the lesion was completely excised from the bone margin without difficulty. The mandibular first molar with root resorption and mobilization was extracted to avoid second dental approach. The crown of the impacted tooth related to the lesion were preserved when the surgical area was curetted. Primary closure of the surgical area was performed with 4/0 Monocyrl. The surgically removed curettage material was sent for histopathologic examination and the patient was scheduled for frequent routine follow-ups.

Histological evaluation

The tissue samples were stained with haematoxylin and eosin. Spindle cells with well-rounded, small nuclei and evidence of myofibroblastic differentiation were observed. Those cells were examined in nodular fasciitis and follicular dendritic tumours. However, they present positive CD21 staining, which is an exception for myofibroma. Morphological findings were compatible with myofibroblastic tumours, while a typical biphasic image was not seen. Additionally, there was a potential for recurrence because of the high proliferative activity of spindle cells (Figure 3). An immunohistochemical analysis including staining for alpha-smooth muscle actin (α-SMA), caldesmon, pHH3, Ki67 and desmin was implemented. The tumour cell staining was negative for desmin and caldesmon, but it was positive for α-SMA, which exhibited a pale colour staining. Furthermore, pHH3 was observed in ten large magnification areas in three cells. A high proliferative rate (< 15%) of Ki67 was identified in the tumour cells (Table 1).

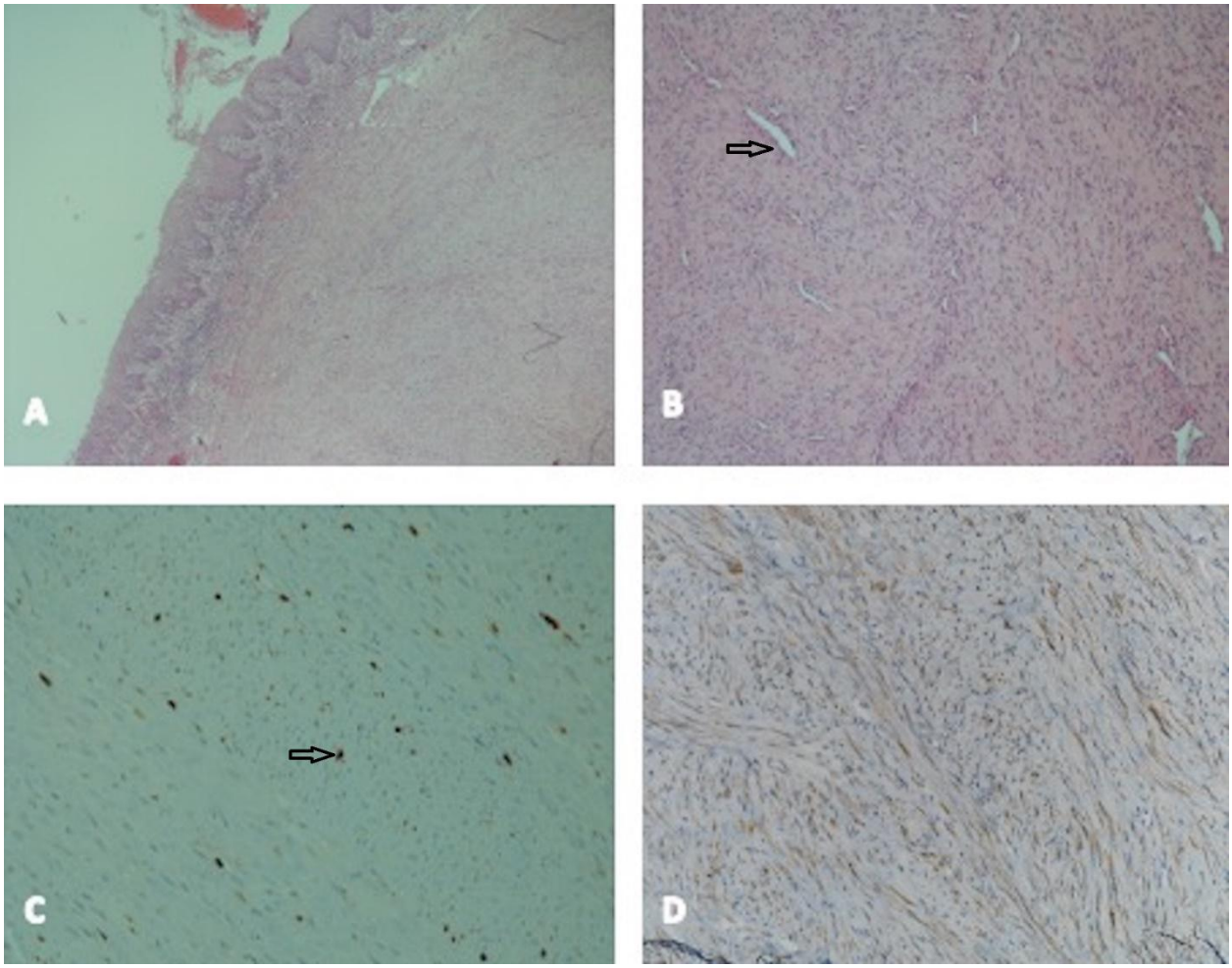

Figure 3 a) Submucosal nodulary lesion (Haematoxylin- eosin, x4). **b)** High power view of the lesion. Note the loosely arranged plump spindled myofibroblastic cells and slit-like vasculary spaces (Haematoxylin-eosin, x10). **c)** Ki-67 proliferation index increased up to %15 in some areas. **d)** Myofibroblastic cells faintly staining with smooth muscle actin (SMA,x10).

Table 1: Immunohistochemical examination after excision

Biomarkers	(+) / (-)
α-SMA	+
Caldesmon	-
PHH3	+
Ki67	+
Desmin	-

RESULTS

Radiologic and clinical examination was performed for 2 years, with a period of every 6 months in the first year and then once a year in the second year. During the follow-up sessions, the lesion showed no signs of recurrence (Figure 4). In addition, soft tissue healing and the permanent teeth, which erupted after the tumour pressure was removed, were indicated clinically (Figure 5).

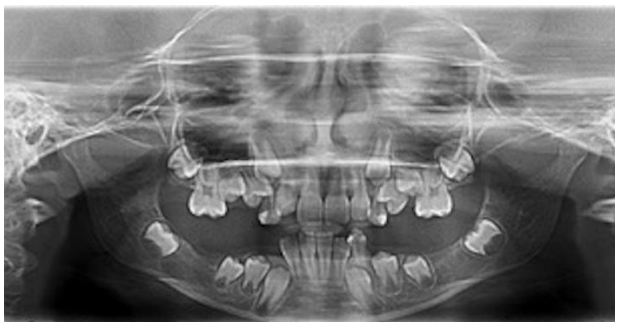

Figure 4: A first year, the bone healing and lack of pressure on the mandibular premolars were observed.

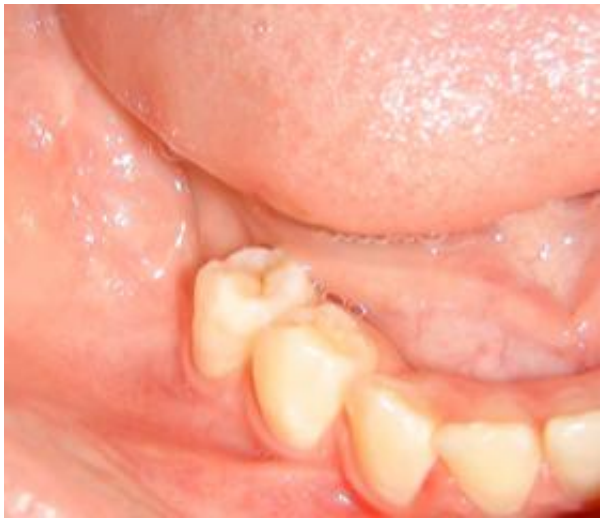

Figure 5: After 2 years, the soft tissue healing and eruption of the mandibular premolars were observed.

DISCUSSION

Myofibroma is a solitary benign tumour that is aggressive and can develop into a rapidly growing mass or an asymptomatic, painless intraosseous mass[2,13]. Myofibroma typically occurs during the first decade of life and is more common in males than in females[5]. Some authors suggest that genetic inheritance with an autosomal dominant or recessive trait could contribute to the formation of this tumour. But the aetiology of this tumour is still unknown[13].

In cases where the jaws are affected, myofibroma manifests as a painless, destructive osteolytic lesion with well-defined borders and slow growth. The clinical signs and symptoms observed are largely contingent upon the specific anatomical site affected, exhibiting considerable variability[14].

Myofibroma represents a rare occurrence within the oral cavity and exhibits morphological similarity with other malignant or benign mesenchymal lesions. This situation can present a challenge in making an accurate diagnosis[15].

Odontogenic lesions such as dentigerous cyst, eosinophilic granuloma, ameloblastoma or nodular fasciitis could have a differential diagnosis of myofibroma clinically[1]. To prevent subsequent deformity and eventual destruction of anatomic structures in children and adolescents, it is essential to ensure an accurate diagnosis and to perform the appropriate surgical intervention. A meticulous histological and immunohistochemical evaluation can prevent erroneous diagnoses and unnecessary radical surgery[16].

Histopathologically, proliferative spindle cells and an aggressive pattern of lesion growth are affected in the differential diagnosis, including inflammatory myofibroblastic tumour, myofibrosarcoma, leiomyoma, neurofibroma, schwannoma, fibrosarcoma, leiomyosarcoma, rabdomyosarcoma, haemangiopericytoma or desmoplastic fibroma[1,3]. In order to differentiate myofibroma from neural and smooth muscle tumours, a panel of antibodies for the detection of desmin, h-caldesmon, s-100, and α-SMA antigens has been proposed[14]. α-SMA is a protein that is commonly used to determine myofibroblastic formation[1-3,5,11,18,19]. The ACTA2 gene codes this protein on 10q22-q24. It is a main factor of contraction. Desmin is a muscle protein that regulates sarcomere structure. It is considered to transmit information of the α-SMA protein. However, caldesmon is a protein that inhibits ATPase activity in smooth muscles[20,21]. In a differential diagnosis, neural tumours, including neurofibroma, neurofibromatosis and schwannoma, are negative for desmin and actin staining. In addition, leiomyoma is a benign smooth muscle tumour that was referred to as a fibroid, and it is positive for desmin, caldesmon and α-SMA staining. Therefore, this tumour could be difficult to distinguish from myofibroma, and it is important that it be assessed clinically as radiologic or oral findings[3,21].

pHH3 is a marker for mitotic activity in early prophase. Staining shows that it is non-specific for neutrophils, macrophages and mast cells. Additionally, Ki67 is a marker that identifies proliferative cells within a tumour. This protein is used to determine an aggressive potential and whether metastasis could result in this tumour. These biomarkers should be considered rather than those of inflammatory myofibroblastic tumour (IMT)[20]. In addition, staining for immunohistochemical

analyses of IMT and alanine aminotransferase (ALT) plays an important role. This enzyme affects the nervous system in 50% of positive IMT cases[20]. However, there is no positive staining for the anaplastic lymphoma kinase (ALK) biomarker in myofibroma.

In the present case, the tumour cells showed negative staining for desmin and caldesmon; however, α-SMA was stained positively with a pale colour. Furthermore, pHH3 was observed in ten large magnification areas in three cells. Ki67 was identified a high proliferative rate (< %15) in the tumour cells.

The number of well-documented cases with long-term follow-up is limited, as is the experience with myofibroma. This situation makes it challenging to determine the best treatment. As is acknowledged, the defining characteristics of myofibroma are well-defined borders, a benign nature, and low recurrence rates. Consequently, conservative surgery with enucleation and/or curettage represents the optimal treatment option[17]. Actually,the patient's age is an effective factor for total or partial radical resection as an en block excision or conservative treatment option[12]. Enucleation or curettage was mostly recognised as the best treatment option[13]. However, Hajeri et al.[2] demonstrated myofibroblastoma in a 3-year-old boy, and after the tumour excision was performed, they preferred to perform reconstruction surgery with a vascularized osteocutaneous fibula flap. In the present case, a minimally invasive approach was preferred in order to prevent the development of long-term complications in a paediatric patient. We performed excisional biopsy, as described by Souza et al.[18], and curettage was carefully executed to protect second permanent molar that were related to the lesion. Clinical and radiographical examination was performed during 2 years of follow-up, leading to no evidence of recurrence.

CONCLUSION

Myofibroma with mandibular bone involvement is rare in children. Conservative management affected the life quality of the patient in this case. We consider that treatment options can be organized to avoid radical resection, although tooth extraction, especially tooth mobility, and bone destruction are decisive factors. Immunohistochemical staining is required to accurate diagnose of myofibroma and identify the nature of tumour cells. Also, follow-up of patients can be preventive to aggressive treatment options even if there is recurrence of myofibroma.

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