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Case Report

An aggressive case of juvenile ossifying fibroma of the craniofacial skeleton

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ABSTRACT

Juvenile ossifying fibroma (JOF) is a rare fibro-osseous neoplasm that develops among the craniofacial bones at an early age. Their aggressiveness added to their high tendency to recur, provoke real diagnostic and therapeutic challenges. We present an agressive case of a nineteen-year-old girl showing facial asymmetry with a large facial mass. The lesion was surgically excised with a satisfactory esthetic result, besides maintenance of the physiological functions. After two years there is no evidence of recurrence of the lesion. Although JOF is a benign lesion, it typically presents aggressive behavior. The high recurrence rates justifies long-term clinical and radiological surveillance.

1. Introduction

Benign fibro-bony lesions in the head and neck region are relatively uncommon entities that cover a broad spectrum of pathologies and share several histopathological features. This group of lesions includes fibrous dysplasia, cemento-osseous dysplasias and ossifying fibroma (ossifying fibroma and juvenile ossifying fibroma) [1].

Juvenile ossifying fibroma (JOF) predominantly affects patients aged under 15 [2]. In most cases, although asymptomatic, this condition presents aggressive clinical behavior, with rapid growth that can provoke extensive facial asymmetry when diagnosed too late. In imaging findings, it is shown as a well delimited uni or multilocular radiolucent lesion, which typically exhibits intralesional radiopaque foci [3]. According to histopathological parameters, the World Health Organization (WHO) considers two JOF variants: trabecular (TJOF) and psammomatoid (PJOF) [4].

The treatment of choice for JOF is surgical excision. According to the clinical presentation of the lesion, the surgical procedure may be conservative or radical [5,6]. Although there are no reports of malignant transformation or metastases, recurrences are observed in about 30%–58% of the cases and, therefore, long-term patient follow-up is indicated [7].

In this context, the present study aims to repor a case of aggressive JOF in the maxilla and discuss clinical, imaging, histopathological and therapeutic findings according to parameters reported in the literature.

2. Case report

A 19-year-old melanoderma female patient presented to a Buccomaxillofacial Surgery and Traumatology service presenting an increase in the volume of the right hemimaxilla, 7.5 cm in diameter, causing facial asymmetry, apparent sclera and deletion of the nasolabial sulcus, presenting 18 months of evolution (Fig. 1A). During intraoral examination, an increase in volume painful to palpation was observed, extending from tooth 12 to 18, approximately 6 cm long, with the oral mucosa presenting normal color (Fig. 1B). The patient was submitted to an incisional biopsy and the histopathological analysis indicated the diagnosis of juvenile ossifying fibroma (Fig. 2).

Computed tomography (CT) revealed the presence of hypodense osteolytic lesions, with intralesional hyperdensal foci, involving the entire right hemimaxilla and extending to the maxillary sinus, right nasal cavity and right orbital floor (Fig. 3A–C).

In order to plan the surgical resection of the tumor, prototypes were prepared to define the lesion resection and to model the titanium mesh to support the resection area (Fig. 3D–F). The Weber Ferguson approach was performed during the surgical approach of the lesion, with exposure of the entire anterior portion of the maxilla, frontal maxilla process, frontozygomatic suture and zygomatic arch. An osteotomy was performed between the lateral and central incisor, nasal fossa region, frontal portion of the maxilla, sphenozygomatic suture, frontozygomatic suture, zygomatic arch and the pterygoid plaque region (Fig. 4A). After total removal of the tumor mass, the surgical defect was reconstructed using the pre-formed titanium mesh (Fig. 4B). The buccal

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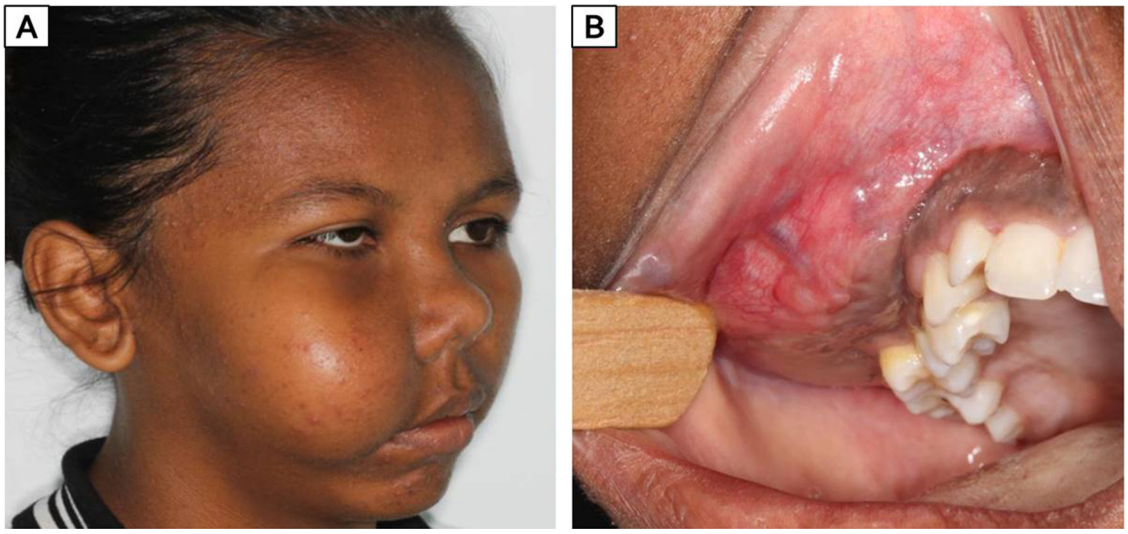

Fig. 1. Clinical aspect (A) Diffuse swelling on right side of the face. (B) Swelling of teeth 12–18, with obliteration of the vestibule.

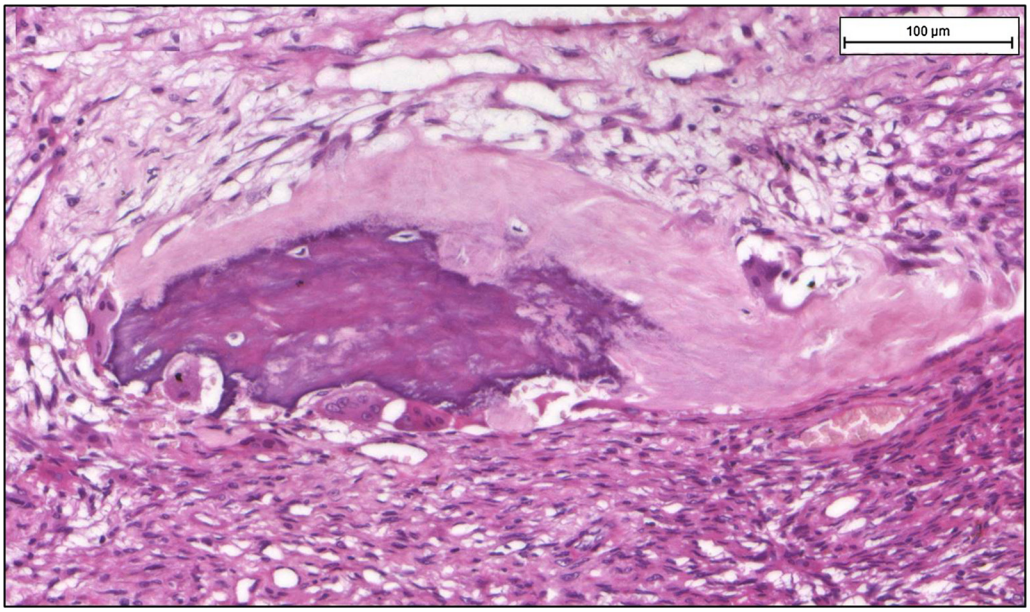

Fig. 2. Histopathological features of incisional biopsy. Mineralized tissue with varying degrees of maturation in a cellular fibrous stroma.

adipose body was exposed and used to assist in the closure of the surgical wound. A transnasal moorage was also used in order to guarantee the positioning of the lateral corner of the eye, avoiding the appearance of hypertelorism (Fig. 4C).

The histopathological analysis of the removed specimens revealed the presence of a benign neoplasia of bone origin, characterized by the presence of irregular bone trabeculae at varying degrees of maturation, with osteoblastic paving and innumerable osteoclasts. The tumor stroma was composed of fibrous connective tissue of variable density and proliferating fusiform and ovoid mesenchymal cells, as well as extensive foci of giant multinucleated cells with different nuclei size and numbers (Fig. 5A–C). From these findings, the JOF diagnosis, previously established in the incisional biopsy, was confirmed.

A panoramic radiography after 30 days of postoperative control evidenced the stability of the titanium mesh (Fig. 6A). The patient has been in the postoperative period for two years. A satisfactory facial aesthetic is observed, as well as maintenance of masticatory, respiratory and ophthalmological functions (Fig. 6B).

3. Discussion

JOF represents about 2% of tumors presenting in the maxillofacial complex. Although no etiology has yet been fully elucidated, studies suggest that non-random chromosome breaks in Xq26 and 2q33 result in a translocation (X;2) that may be associated to the origin of this neoplasm [8].

JOF appears at an early age, with approximately 79% of cases diagnosed before 15 years of age, with no significant gender preference [9]. However, JOF reports in patients aged 3 months to 72 years old have been described [4]. The present study describes a case in a female patient, aged 19. As observed herein, this lesion presents aggressive clinical behavior and rapid growth, capable of reaching considerable dimensions, causing facial asymmetry, with the maxilla being the most commonly affected anatomical site [10].

CT represents an important tool for the diagnosis and surgical planning of JOF cases. Although an aggressive biological behavior is noted, this tumor is typically well defined, which provides an important subsidy for the differential diagnosis of fibrous dysplasia, since that