Odontogenic Myxofibroma in a Paediatric Patient: Case Report and Review of the Literature

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Abstract: This is a case report of an Odontogenic myxofibroma in a paediatric patient. This lesion, also called odontogenic myxoma, is an uncommon benign odontogenic tumour that derives from mesenchymal elements. Odontogenic myxofibroma in paediatric age is rare, because is more frequently discovered between the second and the fourth decade. Clinical, radiological and histological examinations were performed. Clinical examination showed a 3 cm painful swelling in the anterior mandible, and the panoramic radiograph confirmed the presence of an ovoid, well-defined radiotransparent area. Histologically, the odontogenic myxofibroma was characterized by an abundant mucoid stroma containing angular, spindle-shaped and rounded cells. The young age of the patient was considered an indication to perform a conservative approach, with the goal of avoiding the extraction of the elements interested. As reported in literature, the follow-up management depends on several variables, such as location, size, age of onset, and the type of surgical treatment. After 5 years, clinical and radiographic follow up showed no recurrence and the integrity of the anterior mandibular teeth.

Keywords: Odontogenic myxofibroma, Paediatric patient, Odontogenic tumor.

1. INTRODUCTION

Odontogenic myxofibroma (OM), also called odontogenic myxoma, is an uncommon benign lesion of the head and neck that derives from dental embryonic mesenchymal elements [1], which shows a locally invasive behaviour. According to the World Health Organization (WHO), the odontogenic myxoma is classified as a benign tumor of ectomesenchymal origin, with or without odontogenic epithelium [1]. The OM is an lesion that occurs primarily in adulthood, particularly during III decade, while it is rare in children [2]. Some authors have reported that about 60% of OM are located in the mandible, especially in the premolar-molar region [3], while most of the authors have described a uniform distribution of the tumor in the jaw bones [1]. Regarding the frequency of this lesion, Meleti et al. reported an incidence of approximately 0.05 new cases per million population per year [4]. The elevated growth potential, the tendency to local infiltration and different onset sites make the OM a lesion with large clinical and radiological variability.

Clinically, the OM presents painless, invasive, and can cause a marked asymmetry of the face. Radiographically its appearance varies from a unilocular radiolucency with a trabecular pattern trabeculated look at soap bubbles, a tennis racket or honeycomb. Histologically, the OM is characterized by an abundant mucoid stroma containing angular, spindle-shaped and rounded cells. Cellular and nuclear pleomorphism are as well as mitosis. The stroma is usually relatively avascular or may pose delicate capillaries [5].

Recent studies on the biology of neoplastic progression point out the presence of “basal” mechanisms that could assure growth vantages to neoplastic cells. In fact, these cells showed an increased number of mitosis and/or a reduction of apoptosis process.

The treatment of OM includes conservative methods such as local excision, enucleation and curettage, and invasive methods such as en bloc resection. There are several factors that influence surgical choices, such as the location, size, and age of the patient. Recurrence is probably caused by local invasion of bone, and seems to be related to the type of therapy: with conservative surgery these are more frequent [4]. Herein, we reported the feasibility of a conservative approach in the surgical treatment of a new case of OM in a young patient.

2. CASE REPORT

In September 2009, a 11-years-old Caucasian male, in good general health conditions, was referred to the Department of Dental Sciences and Surgery, University of Bari, with a 2-month history of intraoral swelling in the chin of the jaw. The family history did not indicate...
any similar or related anomalies. No previous trauma in the region of the face or mouth was recalled.

Intraoral examination showed a 3 cm-diameter painful swelling in the inferior incisive region that caused the vestibular displacement and rotation of dental elements. Furthermore, the tumour growth led to the loss of vitality of the mandibular central incisors.

A panoramic radiograph was performed. It showed the presence of an ovoid, well-defined radiotransparent area localized in the area of the inferior incisives with a diameter of about 3 cm (Figure 1). Radiographically, the borders of the lesion interested the roots of the inferior incisive teeth. The lesion appeared unilocular for the absence of bony septa. Vitality tests for the central incisors were negative, while the lateral incisors were found to be vital.

Considering the young age of the patient, a conservative approach that avoided the extraction of the elements interested by the lesion was chosen. Therefore, after the endodontic treatment of the central inferior incisors, the tumor was surgically removed with enucleation with peripheral ostectomy under general anaesthesia. At the same time, apicectomy of the left inferior central incisive was performed. Intraoperative examination of the lesion revealed the lack of a perilesional capsule and therefore of a useful cleavage plan. Hence, an accurate instrumental curettage with rotating tools of the residual cavity and application of Carnoy’s solution on the edge of remaining bone were carried out.

The excised material was sent for histopathological examination. Gross examination of the lesion showed a greyish mass measuring 3 X 3.5 X 1.5 cm. Histological evaluation showed randomly oriented spindle-shaped and stellate cells with long cytoplasmic processes, dispersed in mucoid stroma, and no type of calcification has been identified. Haematoxylin-eosin and immunohistochemical staining for vimentin showed features consistent with a diagnosis of OM (Figure 2a-b). No

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Figure 1. An OPT showing the presence of an ovoid, unilocular, definite radiotransparent area, interesting the roots of the inferior incisors, with a diameter of about 3 cm.

Figure 2. Histological features of OM 2A): OM with low cellularity, formed by round, angular and star shaped cells vimentin-positive (Vimentin x 250); 2B): The typical randomly-oriented OM cells are surrounded by a stroma of mucoid substance (Alcian Blue x 100).
bone grafts or allograft materials for filling the residual bone defects were used. The 5-year clinical and radiographic follow up (Figure 3) showed no recurrence and the integrity of the teeth involved in the expansion process of the tumour.

3. DISCUSSION

The OM is benign but locally aggressive odontogenic tumor that can occur at any age, but is more frequently discovered after the second decade. In fact, in 60% to 75% of cases, OM is diagnosed in the second or third decade of life, making unusual the appearance of OM in paediatric patients. Several authors have reported cases of pediatric OM in the literature. The most significant cases, that we found, are reported in Table 1.

Up to 1990 Leiberman et al. were able to document only 17 cases of myxomas in the maxilla in patients aged 14 years or less [6]. Regarding the early childhood, Kadleb et al. have described 21 cases as a separate entity, defining the Infant Odontogenic Myxoma, emphasizing the relatively homogeneous characteristics of this subgroup of patients. In fact, these patients shared several features, such as the age of onset, the localization in paranasal region, and histological appearances [7]. As reported in a recent review article by Meleti et al., the mandible is significantly more affected than maxilla, especially in the posterior region, while the age of onset shows a peak frequency during the third decade [4]. In this report, we present an atypical case of OM, both for tumour location and age of onset.

Regarding the clinical features of this tumour, the majority of OM are asymptomatic, probably due to the fact that many patients have a tumour in the posterior regions, and therefore they are less disfiguring [4]. Because the odontogenic tumours have overlapping clinical and radiological features, the definitive diagnosis of OM is based on histopathological examination.

As previously reported, OM presents stellate, spindle-shaped and round cells combined through their cellular processes, dispersed in mucoid or myxoid stroma, with a variable amount of collagen fibers. Regarding rests of odontogenic epithelium, they are not required for diagnosis [8].

At histological level, there are several tumours that can resemble OM, such as odontogenic fibroma, cemento-ossifying fibroma, chondromyxoma tumours, desmoplastic fibroma and fibroosseous lesions, with problems in differential diagnosis [9,10].

Odontogenic and cemento-ossifying fibroma are densely collagenized, and may presents dentinoid or osteoid and cementum areas more or less extensively disseminated [3,11]. Chondromyxoma, although can have a myxoid stroma, they also presents catilagenous areas, while desmoplastic fibroma have a greater degree of cellularity and fascicles of collagen fibers [10]. Microscopically, the current case showed a tissue with low cellularity, containing round, angular and star-shaped cells vimentin-positive, surrounded by a stroma of mucoid substance.

Radiographically, it may produce several patterns: unilocular, multilocular, or with less frequent patterns, such as pericoronal or radiolucent–radiopaque. This great variability and the lack of specificity of radiological signs makes mandatory histopathological examination, due to the board range of causes, both of odontogenic and non odontogenic origin [12].

Figure 3. 5-years radiographic follow up showing the good healing in the area previously interested by the lesion.
Radiographic features of OM are the presence of a unilocular or multilocular radiolucency with a honeycomb. From the data present in the literature, multilocular pattern seems to be slightly more common [4,13]. The present case showed an ovoid, unilocular, definite radiotransparent area, interesting the inferior incisive roots. Displacement of the teeth can be a characteristic of odontogenic tumours, and specifically of OM, when they reaches a certain size, as in the present case.
Treatments options range from conservative approach, which consists on enucleation of the tumour and curettage, to the complete resection of the lesion including wide bone margins, to avoid recurrences. According to several authors, a conservative approach is preferred in the cases affecting mandible, while a radical approach is preferred in maxillo-orbital region, due to the fact that any OM recurrences would affect a functionally and aesthetically critical area. Furthermore, no evidence of malignant degeneration of OM was reported in literature [4,13,14]. In the present case, considering the young age of the patient, a major goal of the treatment was to avoid the extraction of the elements interested by the lesion.

On the other hand, due to its tendency to local infiltration and difficulty of finding a plan of cleavage, OM recurrences are frequently reported [15]. For this reason, patients should be followed indefinitely, but especially in the first 2 years after the surgical treatment, because the majority of recurrences of OM reported in literature tend to appear early (Table 1) [13,16]. In this case, the follow-up continued for 5 years, confirming the absence of recurrence.

From the data obtained it is not possible to determine the specific characteristics of OM in children and adolescents, as has been done for infants; however, the number of cases reported makes us think that this rare odontogenic neoplasm must still be considered in the differential diagnosis of radiolucent lesions of both jaws in all age groups. Addition it is not possible estimating the possibility of recurrences with conservative surgery in children and adolescent patient because many of the cases examined did not specify the treatment, follow-up and recurrence, but in adult patient recurrences are most common with conservative surgery.

Herein, we illustrated a case of OM in a young patient that reached a complete healing performing a conservative surgical approach. The disappearance of the lesion, the absence of relapse in a 5-year follow-up period and the recovery of the dental elements were the goals achieved.

To date, it is not possible to determine what is the most suitable approach in cases of OM involving paediatric patients because of the paucity of similar cases in the literature. In this case, conservative treatment has been successful. If the lesion involves dental apex, an endodontic treatment and subsequent apexectomy should be the choice option instead of dental extraction. However, further cases are needed to confirm these issues.

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