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Up-to Date Review And Case Report

Case report of maxillary osteosarcoma with up-to date review

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Abstract – Introduction: Osteosarcoma is characterized by its rarity and malignant nature, leading to difficulty in diagnosis. **Observation:** We present the case of a young woman observed by her orthodontist for a recent asymptomatic intraoral mass. The first anatomo-pathological result of the sample was a benign lesion, but rapid recurrence of the lesion led to the samples being retested, resulting in the diagnosis of osteosarcoma. **Discussion:** The most common symptom associated with osteosarcoma is painless swelling, sometimes associated with dysesthesia and/or limited mouth opening. The radiological signs may be of the osteogenic- or osteolytic-type. After histological confirmation the treatment of this tumor requires surgical excision sometimes combined with chemotherapy. **Conclusion:** Osteosarcoma should be part of the differential diagnosis when a patient has an intraoral bone tumor.

Introduction

Osteosarcoma, or osteogenic sarcoma, is a malignant bone tumor in which neoplastic cells produce osteoid bone or immature bone. Osteosarcoma of the jaw is rare and accounts for approximately 7% of all osteosarcomas. Compared to the clinical presentation of osteosarcoma of the long bones, osteosarcoma of the jaw occurs in older patient population (30–50 years). The mandible is more often affected than the maxilla [1]. These rare tumors are of mesenchymal origin; osteosarcoma and chondrosarcoma are the most common histological types followed by Ewing's sarcoma [2]. During the clinical examination, pain and swelling of the face and trismus might be observed [3]. Given the rarity of this pathology and the lack of specific signs, the diagnosis is often difficult and requires a multidisciplinary discussion.

Herein, we describe the case of a patient who presented to the oral surgery department.

Observation

A 17-year-old patient was referred for oral surgery for a recent dental displacement accompanied by a gingival arch. The clinical examination revealed a vestibular arch between the right maxillary lateral incisor (tooth 12) and the right maxillary canine (tooth 13). This arch was of hard consistency, immobile, and covered by a healthy mucous membrane (Fig. 1). Three-dimensional imaging was performed to support the diagnosis (Fig. 2). A heterogeneous radio-opaque mass was observed between tooth 12 and 13 without rupture of the cortex.

According to the usual procedure followed upon the appearance of a suspicious lesion, we decided to perform excision biopsy of this anomaly using a piezotome. The sample was then sent to the pathology lab (Fig. 3). Histological analysis (Figs. 4 and 5) did not show any microscopic evidence in favor of malignancy; therefore, the diagnosis of osteochondroma was made.

Unfortunately, 2 months later, the patient had a recurrence of the lesion. After a multidisciplinary discussion, a maxillofacial surgeon and a pathologist suggested retesting of the samples. The first hypothesis put forward was the diagnosis of fibrous epulis with bony and more cartilaginous metaplasia. Considering that it was a gingival lesion and knowing that the deepest extent of the lesion passed into the lesion area (Fig. 4), this pathology seemed possible. However, an expert pathologist re-examined the specimen's histology and made a diagnosis of conventional high-grade malignant osteosarcoma that was predominantly osteoblastic with a chondroblastic and fibroblastic contingent.

The patient was then referred to the pediatric hematology department. She initially received chemotherapy comprising methotrexate (Imeth®) and dexamethasone (Dectancyl®). In a second step, a procedure was performed including a right enterolateral partial maxillectomy with the intraoral excision of the infrastructure of teeth 22 to 16 and reconstruction using a

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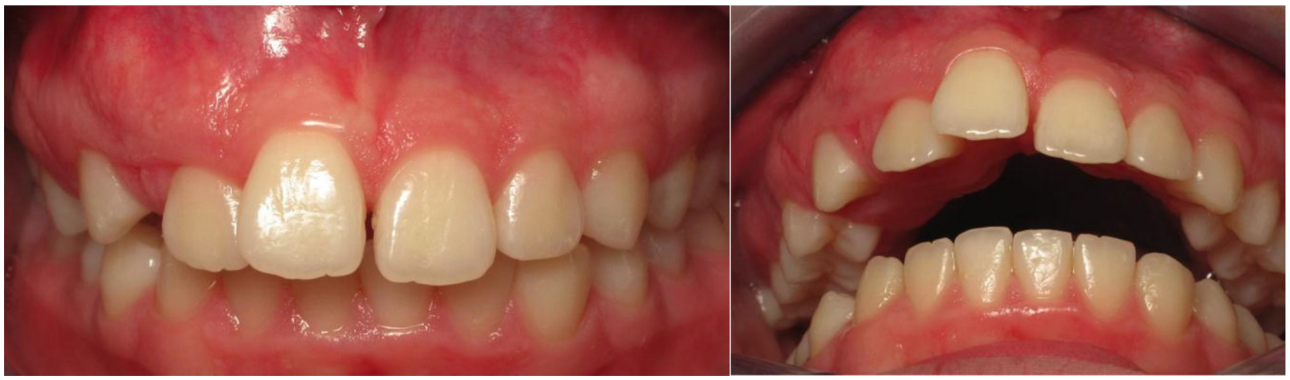

Fig. 1. Gingival vestibular and palatal tumor located between the maxillary right lateral incisor and the right maxillary canine with a homogeneous rosy mucosa, without ulceration.

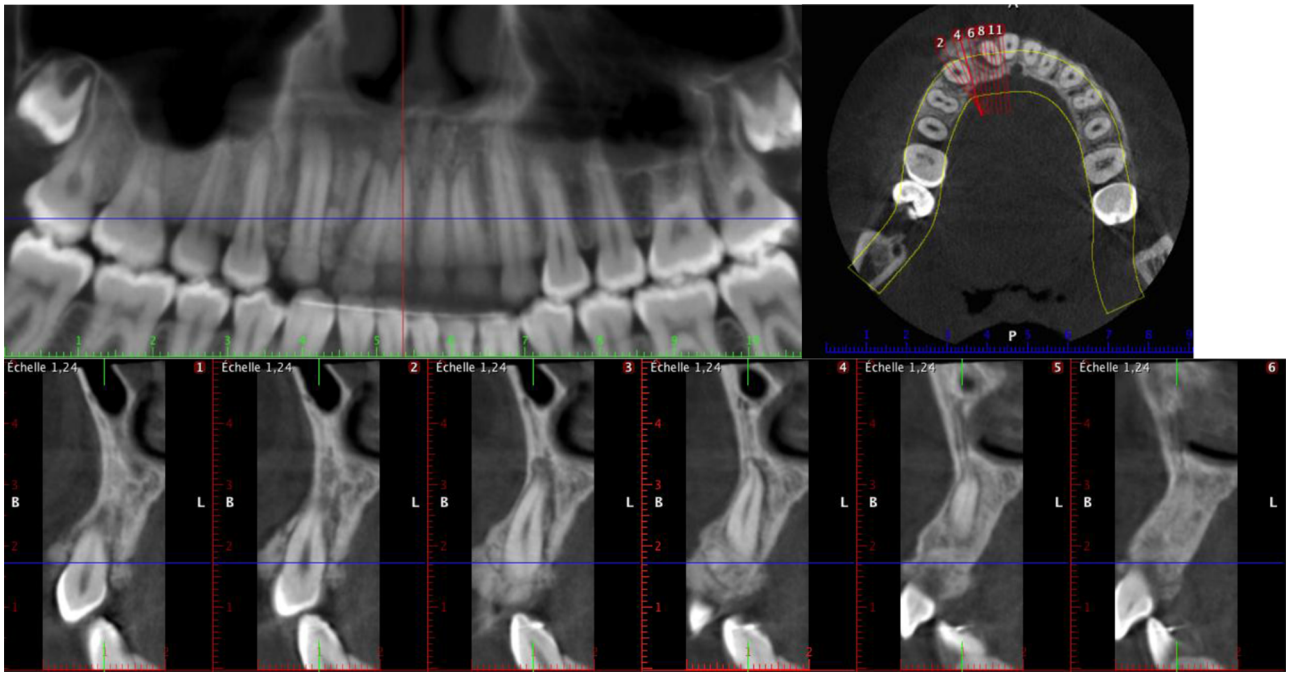

Fig. 2. Heterogeneous image between 12 and 13 with vestibular and palatal involvement and periosteal reaction.

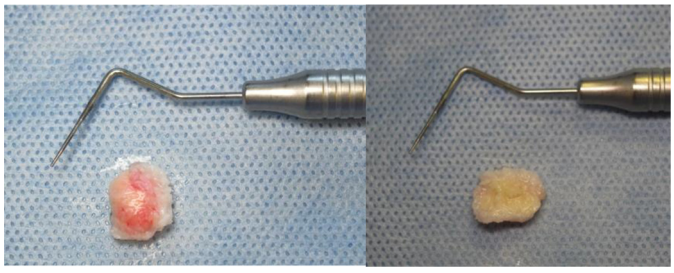

Fig. 3. External surface of the anatomical part with a bony aspect and Intern surface of the anatomical part with a cartilaginous appearance.

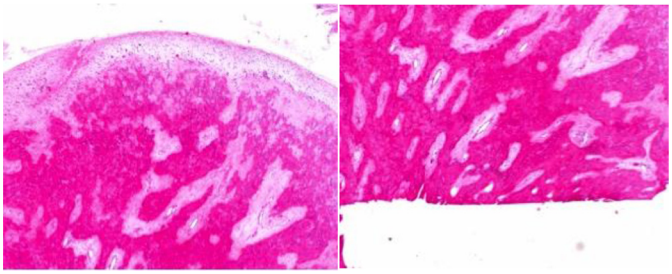

Fig. 4. Views of the upper part of the lesion as well as the deep part. It is a lesion of the bone nature, consisting of a fairly dense fibrous bone, persisted spaces occupied by a highly collagenized tissue, this bone tissue has many resorption stalls. The deep part goes into the lesion area.

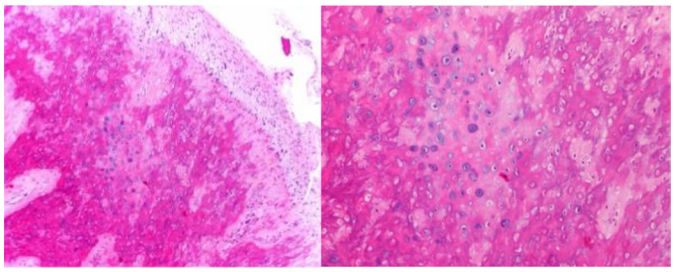

Fig. 5. Section of the upper part of the lesion showing chondroid differentiation.

free flap from the right fibula. Following this second intervention, a second chemotherapy was implemented with adriamycin (Doxorubicine®) and cisplatin (Cisplatin®).

The end-of-treatment assessments have not indicated any anomalies to date.

Discussion

Osteosarcoma of the jaw is difficult to diagnose and manage. The reason for this is the high frequency of errors in biopsy results due to the rare specific radiological characteristics and difficulties in appropriate resections because of the proximity of the vital structures [4].

Confirmation of the final diagnosis of osteosarcoma often requires histopathological examination of multiple biopsy samples and live discussions among experts [5]. The incidence of false biopsy results in bone tumors ranges from 17% to 25% [4].

Maxillary osteosarcomas usually develop during the ages of 20 and 40 years. Osteosarcoma has no specific clinical symptom; it presents with the same clinical signs as all maxillary tumors [6].

The most common complaint is painless swelling, which can sometimes be associated with dysesthesia and/or limited mouth opening [7]. Due to the anatomical location of mandibular osteosarcomas, dentists are the first to assess the lesion in 45% cases. Unfortunately, due to misdiagnosis, tooth extraction occurs in two third of cases and half of the patients are treated with antibiotics [8]. If osteosarcoma of the jaws is suspected, conventional radiographic studies and tomography and/or magnetic resonance imaging are recommended [9].

In osteosarcomas, the radiological characteristics depend on the tumor behavior in the form of bone destruction and formation. They can range from purely osteogenic (appearance of sunbeams) to purely osteolytic or a mixture of both [4]. The enlargement of the periodontal ligament space and the attenuation of the hard blade around a tumor are other characteristics that can appear in osteosarcomas of the jaw [4]. Because this tumor grows very quickly, new bone formation tends to occur in a straight line, at an angle of 90° to the new bone surface [10]. Therefore, the most common X-rays taken by a dentist would miss this, especially during the early stages of the lesion. A second 90° view, such as a true lower occlusal view, is necessary to demonstrate this significant radiographic characteristic.

According to Bianchi and Boccardi, there are the following three categories of appearances on the scan: 1. Radiotransparent with the absence of bone formation in the tumor; 2. Marbled with small areas of amorphous ossification; and 3. "Lamellar" ossification with radiating bony plates from a focus-like ray of sunshine [11].

MRI is important to assess the exact location of the mandibular canal before planning resection of a tumor as well as for reconstruction.

Diagnostic ultrasound can play a central role in the identification of pathological signs, namely the soft tissue mass associated with bone thinning, erosion, dilation, and the "sunbeam" aspect of the oral cortex, suggestive of osteosarcoma [10].

The combination of radiological signs is an important finding that should be recognized but should not be confused with common infectious dental diseases.

Resorption of the teeth is not usually an early characteristic of osteosarcoma because the teeth are highly mineralized and take time to reabsorb. Due to the rapid growth of the tumor, dental resorption does not manifest until the later growth stages of the tumor [10].

A study by Gomez-Brouchet *et al.* demonstrated the effectiveness of the estimation of galactin-1 expression during the differentiation of the chondroblastic variant of osteosarcoma from chondrosarcoma. Galactin-1 is abundant in osteosarcoma but not in chondrosarcoma with a positive predictive value of 85.7% [4].

The negative prognostic factors are as follows: the maxillary localization due to limited margins of excision, tumor size, and osteoblastic subtype [12]. Females with a predominantly chondroblastic pattern may have a poorer prognosis [13].

The essential histological characteristic of osteosarcomas is the existence within a sarcomatoid stroma of skeletal tissue arranged in an anarchic manner and synthesized by atypical osteoblasts, having malignant characteristics [14].

Well-differentiated osteosarcomas present a challenge for histological analysis because they are often confused with benign fibro-osseous lesions such as fibrous dysplasia or ossifying fibroma. These two lesions have irregularly anastomosed and curved bone spans as well as little cytological atypia, on microscopic examination [13].

The treatment of choice for most bone sarcomas, regardless of the histological subtype, is complete surgical resection with wide margins with or without preoperative chemotherapy. A broad resection with clear margins can be achieved at the extremities, but it is difficult in the head and neck regions due to the complex anatomy and proximity to the vital organs, often resulting in incomplete resection leading to a local recurrence. Therefore, surgical resection is often followed by adjuvant radiation therapy and/or chemotherapy to prevent recurrences [2].

Conclusion

In presence of a bone tumor, eliminating the diagnosis of a malignant pathology must remain the priority of practitioners. Rapid management is necessary in these serious pathologies. When in doubt, it is desirable that the pathological results be confirmed by an expert institution.

Conflicts of interests: The authors declare that they have no conflicts of interest in relation to this article.

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