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CASE REPORT

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Undifferentiated osteosarcoma of the mandible oblique line: a case report

Osteossarcoma indiferenciado da mandíbula: relato de caso clínico na região do trígono retromolar

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ABSTRACT

This article reports a case of 82 years old woman, who consulted in a private radiology dental clinic for implants planning. In a panoramic radiography, a radiopaque area with sun-ray appearance located above the external oblique line on the right side was observed. The incisional biopsy showed presence of sarcomatous stroma presenting osteoid tissue with irregularly shaped and large amount of osteoblasts, varied sizes and shapes, with prominent nuclei, intensely colored, arranged in a disorderly way around trabecular bone. Histopathological diagnosis was obtained for undifferentiated osteosarcoma. Osteosarcoma located in the jaw (JOS) is rare and aggressive, constituting 5% to 13% of all cases of skeletal osteosarcoma. JOS has a male predilection in third decade of life.

KEYWORDS

Osteosarcoma; Jaw; Mandible.

RESUMO

O osteossarcoma (OS) é o tumor maligno primário mais comum. Entretanto, o osteossarcoma localizado na mandíbula (JOS) é raro, agressivo e maligno, constituindo de 5-13% dos casos de OS do corpo todo. O JOS tem predileção pelo sexo masculino com idade de 34-36 anos. O prognóstico da doença está associado com diversas variáveis tais como a localização do tumor, fase de desenvolvimento, existência ou não de metástases, sexo, idade, e resposta do organismo ao tratamento. Este artigo relata um caso incomum de osteossarcoma localizado na região do trígono retromolar mandibular, composto por osso mesenquimal primitivo, que comumente ocorre nas regiões de extremidades e ossos longos.

PALAVRAS-CHAVE

Osteossarcoma; Maxilares; Mandíbula.

INTRODUCTION

According to World Health Organization (WHO), osteosarcoma (OS) is the most common primary malignant tumor of bone, accounting for approximately 35 percent of cases, followed by chondrosarcoma (25%), and Ewing sarcoma (16%), and occurs predominantly in patients younger than age of twenty, and in this group 80% occur in long bones of the extremities [1].

Osteosarcoma of the jaw (JOS) is a rare, aggressive malignancy constituting 5% to 13% of all cases of skeletal OS [2]. The signs and symptoms of JOS include pain, paresthesia, regional swelling and the patients can report loose teeth, changes of teeth position or changes in prosthesis adaptation [3].

OS most often affects the rapidly growing parts of the skeleton; arises in the metaphysis of long bones of the extremities. Bone or osteoid formation within the tumor is characteristic of

an osteosarcoma [4,5]. The common sites are femur, tibia and humerus [5]. Furthermore, OS may affect multiple sites. This entity (multifocal or multicentric osteosarcoma) was first described by Silverman in 1936 [6].

In North America and Europe, the incidence rate for bone sarcomas in males is approximately 0.8 new cases per 100,000 population and year. Somewhat higher incidence rates have been observed for males in Argentina and Brazil 1.5-2 and Israel 1.4 cases [1].

The diagnostic is obtained through the X-ray examinations, Computed Tomography (CT) and pathologic analyses. Panoramic radiography remains the primary means of diagnosis, where the image the "sun ray" shows pathognomonic signal, although CT images provide high quality and excellent anatomic resolution, providing visualization of tumor calcification and cortical bone involvement, being of great importance in the diagnostic and treatment planning. The emergence of this disease at the trigonoretromolar region (oblique line) is the novelty of this clinical case. In the most cases the growth of the lesion is low.

Prognostic is associated with several variables, such as tumor location, initial size, existence or absence of metastasis, gender, age, cytogenetic chances and response to chemotherapy pre surgery [7]. According August et al. [8], local recurrence was the most common cause of death in JOS. In their study the average recurrence time was 18 months with a median of 7 months and a range of 0-83 months.

CASE REPORT

Caucasian patient, 82 years old, female consulted a private dental radiology clinic to perform radiographic documentation for placement of implants. During anamnesis reported difficult open the mouth. In the clinical evaluation it was detected a discrete area of fibrous consistency, asymptomatic, with no changes in color and volume in the oral mucosa, located in the right retromolar triangle (Figure 1). Radiographic examination showed radiopaque area with a sun-ray appearance located above the external oblique line on the right side (Figure 2).

Multislice CT scans revealed hyper dense area radiating from the crest of the alveolar ridge in the region of third lower molar on the right side, region of the external oblique line (Figure 3). An incisional biopsy was performed intraoral and showed the presence of sarcomatous stroma presenting osteoid tissue with irregularly shaped and large amount of osteoblasts, varied sizes and shapes, with prominent nuclei, intensely colored, arranged in a disorderly way around trabecular bone (Figure 4). Histopathological diagnosis: undifferentiated osteosarcoma of the mandible.

The radiographic differential diagnosis for JOS includes fibrous dysplasia, osteomyelitis, osteoma, myositis ossificans and cement-osseous dysplasias, they all commonly arise in mandible [9].

The patient was referred to head and neck surgery service where it was held M-section of the mandible (Figure 5 and Figure 6), 10 sessions of chemotherapy and 20 sessions radiotherapy.

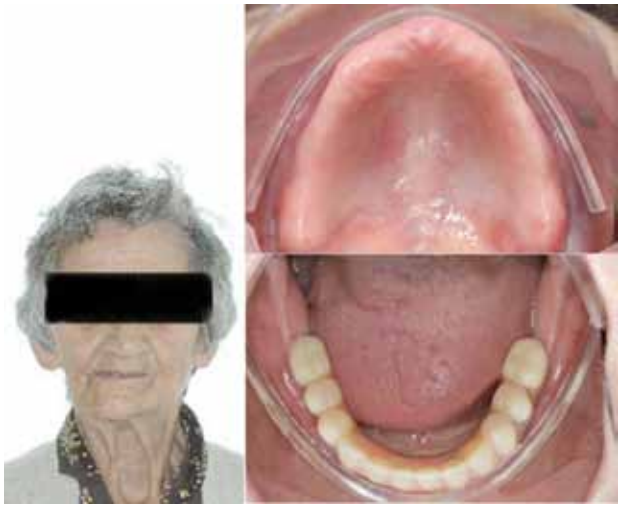

Figure 1 - View the patient's intra e extra –oral before the treatment.

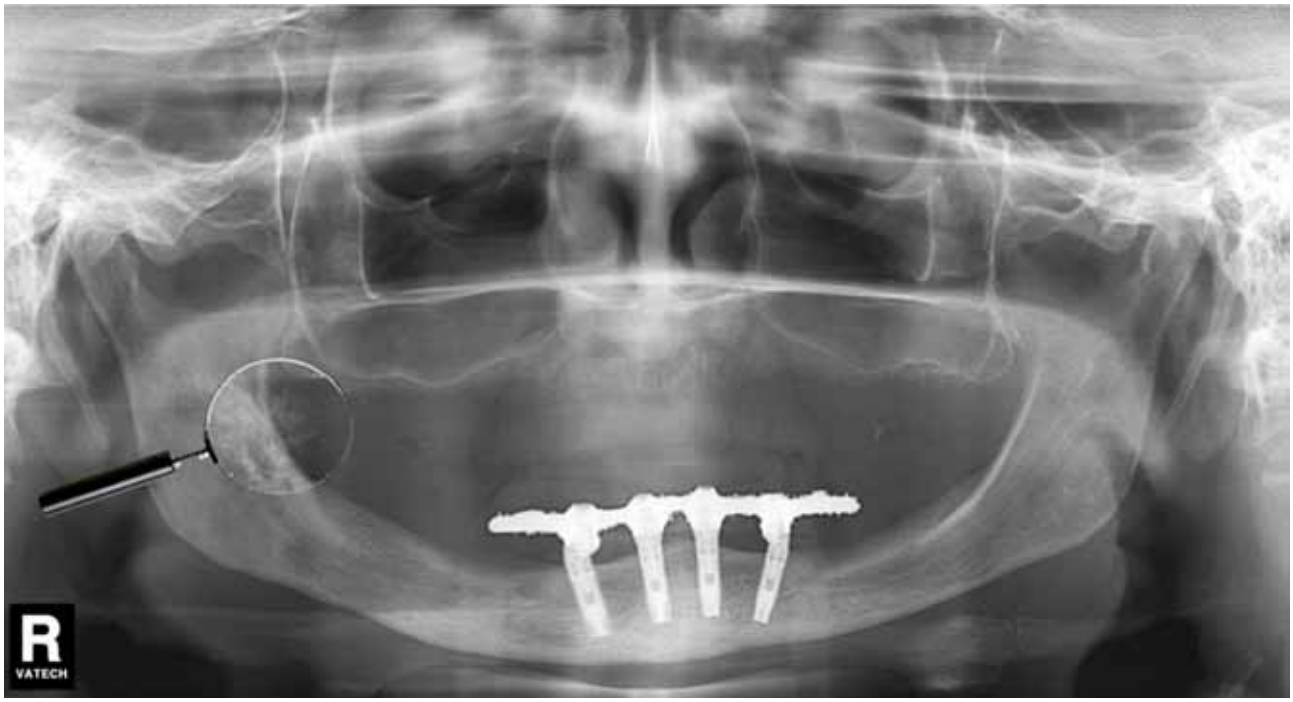

Figure 2 - Panoramic radiograph showing the radiopaque lesion with a sun-ray appearance located above the external oblique line on the right side.

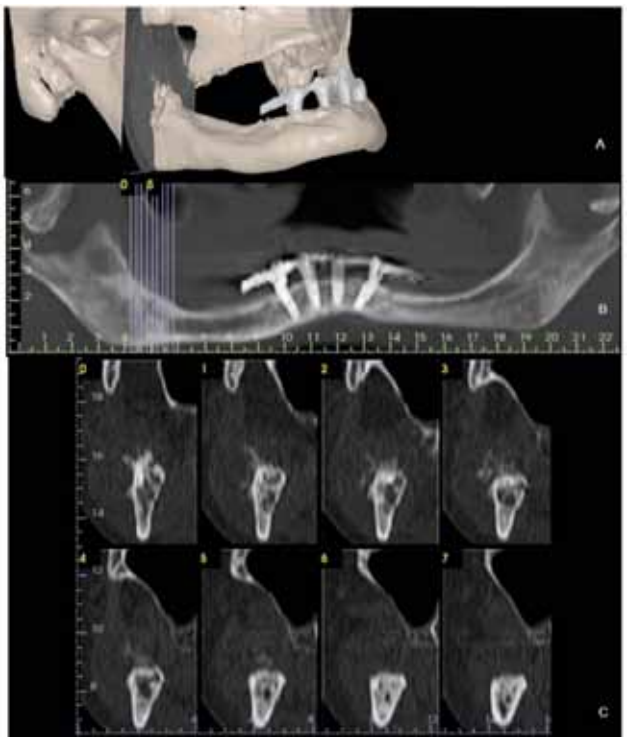

Figure 3 - A) 3D Reconstruction, (B) Panoramic CT with cuts (mm) in region of the external oblique line and (C) Paraxial cuts in full size (1:1), revealed hyper dense area radiating from the crest of the alveolar ridge in the region of third lower molar on the right side.

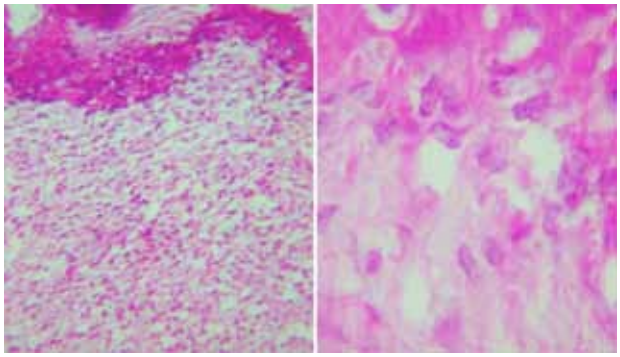

Figure 4 - Microscopy showed the presence of sarcomatous stroma presenting osteoid tissue with irregularly shaped and large amount of osteoblasts, varied sizes and shapes, with prominent nuclei, intensely colored, arranged in a disorderly way around trabecular bone.

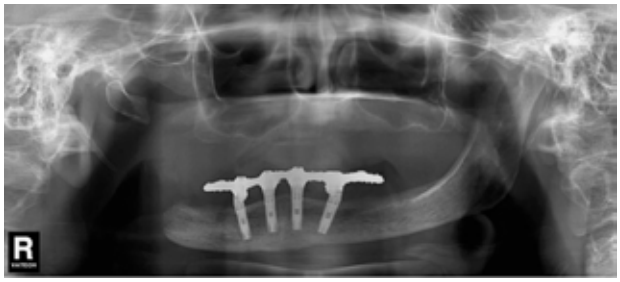

Figure 5 - Panoramic radiograph after removal of the lesion on the right side.

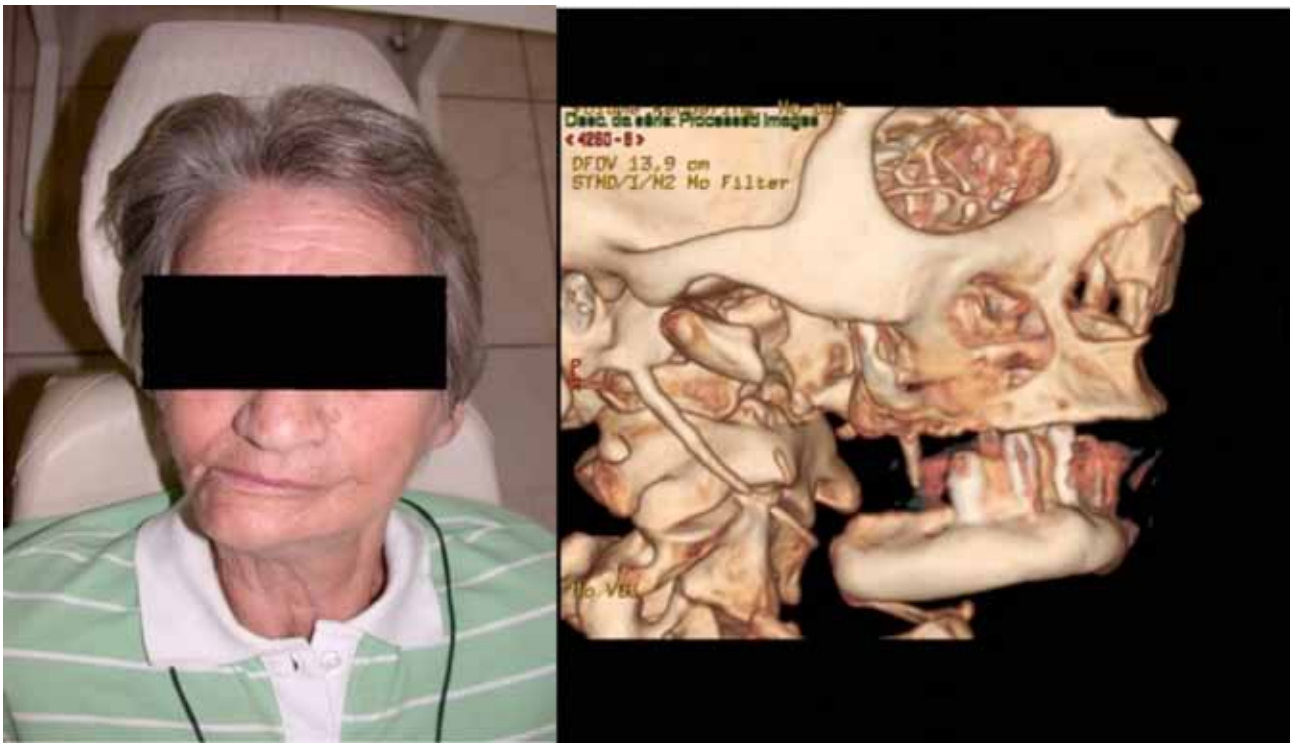

Figure 6 - Oral view of the patient and 3D CT reconstruction after surgery.

DISCUSSION

Osteosarcoma is the most common malignancy of bone, with an age-related incidence of up to 16 per million at the age of 16 years [10]. Most osteosarcomas of the jaw – JOS (87%) are high-grade lesions, while low-grade lesions are rare and include the well-differentiated intramedullary osteosarcoma (or JOS) and the periosteal osteosarcoma. Low-grade central osteosarcoma occurs within a broad age range (16–69 years), with peak incidence in the third or fourth decades [4]. JOS represent of 1 to 9% of the total number of osteosarcomas are mentioned [11] and according to WHO, are very rare, with an incidence of 0.7 per million. They are extremely rare in other head and neck sites [12].

The important point in this case report is the fact the lesion was a radiographic finding. Despite the patient reported difficult open the mouth, she showed no symptomatology described previously. Another point was the sex and age of patient.

The gender distribution of osteosarcoma of the jaw has a male predilection with mean age from 34 to 36 years. The most common clinical characteristic presentation of JOS is swelling with or without numbness and limitation of mouth opening [12].

Nissanka et al. [13] demonstrated the mandibular osteosarcoma is more common than maxillary lesion. Hereditary retinoblastoma, Paget's disease of bone, a history of fibrous dysplasia, or trauma are other factors known to predispose to the development of osteosarcomas [14].

A review of the literature showed 85% to 95.5% of patients with OS has swelling, either painful (45.8% to 50%) or painless (35% to 47.9%). Other complaints included paresthesia (in 21,2% of cases), displacement of teeth,

epistaxis, eye problems, nasal obstruction and weight loss [1].

The lesion found in the panoramic radiograph can generate other diagnostic hypotheses. The fibrous dysplasia involving the jaws, the disease tends to appear early in life. The classic radiographic appearance is a "ground-glass" or "orange peel" effect, a mixed pattern of radiolucency, showing irregular and heavy radiopaque foci; this has been described as a "smoke" pattern [15]. Osteomyelitis of the jaws of a chronic nature has findings consistent with swelling, pain, purulence, intraoral or extra oral draining fistulae; positive radiographic finding is lesion with diffuse sclerosing [16]. Osteoma is a benign osteogenic lesion characterized by the proliferation of compact or cancellous bones, clinically, the peripheral osteoma is usually asymptomatic but can produce swelling and cause asymmetry, radiographically the lesion appears as a well-circumscribed radiopacity [17]. The final diagnosis is made by biopsy and histopathological analysis.

Reports on osteosarcoma of the jaws from Africa are few; most reports originate from Nigeria, Kenya, and South Africa. Ogunlewe et al. [18] reported that osteosarcoma of the jaw constituted 0.6% of lesions of the jaws and oral cavity over a 21-year period in a Nigerian institution; the patients mean age was 27 years. While mandibular lesions occurred predominantly in women, their study suggested a male predisposition of maxillary lesions.

As with a number of other pathologies in sub-Saharan Africa, late presentation of patients with osteosarcoma of the jaws is a common feature. Thus, the use of adjuvant chemo-radiotherapy with surgical extirpation does not appear to affect outcomes, primarily as the result of late presentation [14].

The optimal treatment for jaw metastases of (OS) is radical surgical mandibular resection [13]. Likewise, the most important curative therapy for primary jaw (OS) has been found to be radical resection with clear margins [19].

In this reported case, was performed an m-resection of the mandible. Resection with surgical margins is the most important factor for prognosis and provides a 5 years survival rate of 80% [4]. Bony margins for resection should at least be 2 cm from the clinical radiographic edge or the nearest suture in the mid face. Soft tissue margins around an osteosarcoma resection should be 2 cm or more assessed with frozen section. Adjuvant chemotherapy or radiotherapy seems to be efficacious [20].

The treatment of osteosarcoma of the jaws should be approached in two ways. Radical surgery is the primary treatment for (OS) of long bones as well as jaws, although it cannot be contemplated as the sole treatment. The additional use of radiotherapy was left to the discretion of the treating physician but was generally encourage in cases of incomplete resection [12].

CONCLUSION

Osteosarcoma (OS) is a rare malignance bone tumor of mesenchyme origin. Its development is fast and metastatic. The clinical importance of this case report is that the lesion was a radiographic finding, since the patient had no symptoms, and the diagnosis was made due the indication for placement of implants. The diagnostic is obtained through the x-ray examinations (panoramic, periapical, occlusal), CT and pathologic analyses should be the earliest possible. Prognostic is associated with several variables, such as tumor location, initial size, existence or absence of metastasis, gender, age, cytogenetic chances and respond to chemotherapy pre surgery.

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