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Case Report

Maxillofacial Mazabraud's syndrome: A case report & review

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ABSTRACT

Mazabraud's syndrome is characterized by myxomas of intramuscular type present in association with fibrous dysplasia. Up to this day, approximately 80 cases of Mazabraud's syndrome have been reported, although in the head and neck territory intramuscular myxoma reports in association with fibrous dysplasia of the bone are very scarce. An unusual case of Mazabraud's syndrome in a 63 years old female displaying fibrous dysplasia of the mandible and soft tissue myxoma in the edentulous alveolar ridge in the molar area is reported. After four years of follow-up, the clinical, imagenological and microscopical findings that led to the diagnosis and treatment are discussed. This report exemplifies the diagnostic and treatment challenge of this rare disease and enhances our clinical knowledge due to its long follow-up, highlighting the need of understanding better its behavior in order to establish proper guidelines for its treatment.

1. Introduction

Mazabraud's syndrome (MS) is characterized by myxomas of intramuscular type present in association with fibrous dysplasia [1]. Intramuscular myxoma (IM) is a benign mesenchymal tumor and fibrous dysplasia (FD) is a benign intramedullary fibro-osseous lesion [2].

MS was first depicted by Henschen [2] in 1926 and in 1967 a pattern of association between FD and soft tissue myxomas was described by Mazabraud et al. [3]. Up to this day, approximately 80 cases of Mazabraud's syndrome have been reported [4].

Clinical features usually involve multiple IMs frequently located in lower limbs; typically, myxomas are located adjacent to the bone lesions [5]. Commonly associated with the polyostotic form of FD, but monostotic involvement has been reported as well. FD's onset precedes IMs, usually asymptomatically and frequently involving women [1,5].

Solitary myxoma and monostotic FD is rarely reported, moreover, MS is extremely infrequent in the head and neck area. Here, a case of mandibular Mazabraud's syndrome with a 4-year follow-up and a review of this rare disease are reported.

2. Case report

2.1. History and background

A 63-year-old female patient was referred to the oral and maxillofacial surgery service for assessment of a tumor located in the left mandibular body.

The patient reported the tumor had been present since adolescence. Given the lack of symptoms, no medical opinion was sought. When the patient was 47 years old, surgical removal of the tumor was indicated to craft a denture. The tumor relapsed soon after excision difficulting proper chewing years later.

Regarding her medical history, type 2 diabetes mellitus, hypertension, allergy to lysine clonixinate and a habit of 10 years cigarette smoking was reported.

2.2. Clinical findings

Upon examination, a large sessile tumor of rubbery consistence and 8 centimeters of major diameter was found in the edentulous alveolar ridge in the molar area. Notably asymptomatic, painless on palpation, with lobulated surface and traumatic ulcers related

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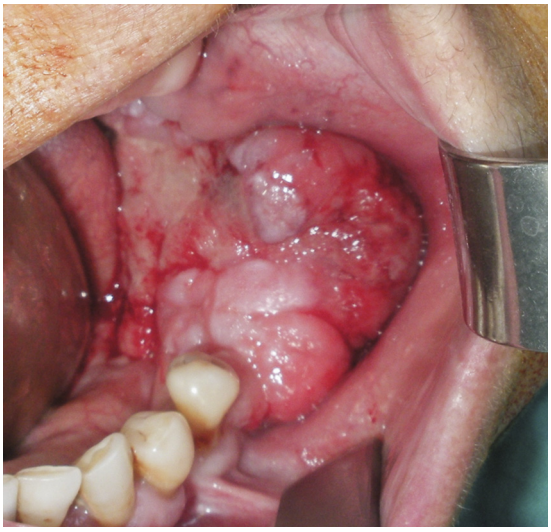

Fig. 1. Tumoral mass at the inner aspect of the left cheek, before excision.

to the occlusal plane of neighboring teeth. No other relevant findings were found (Fig. 1).

Blood samples were requested revealing alkaline phosphatase and glycosylated hemoglobin levels were higher than normal, evidencing poor metabolic control of type 2 diabetes and raising suspicion of endocrine disruption.

2.3. Imagenological findings

Computed tomography (CT) showed an expansive process in the body and ramus of the left mandible with ill-defined borders and thinned cortical bone expanding lingually and buccally. Two different patterns were observed: the body of the mandible showed ground glass appearance (Fig. 2A), while the ramus showed an expansive process with cortical defects at the medial cortical outline (Fig. 2B). Cranial and distal displacement of the third left molar was noted (Fig. 2C). The process was associated with mucosal thickening of undetermined appearance.

It was agreed that a biopsy of the mandibular tumor and subjacent bone were necessary.

2.4. Surgical procedure

An incisional biopsy was performed at the proximal aspect of the intra-oral swelling. The sample was diagnosed as oral submucous fibrosis by a general pathologist.

Given the apparent benign character of the lesion, a second procedure was scheduled, with the aim of fully removing the soft tissue lesion and to perform an incisional biopsy of the subjacent bone. The full excision of the mass was successfully achieved (Fig. 3A). An incisional biopsy of approximately 2 centimeters was taken immediately distal to the second lower bicuspid comprising cortical bone and marrow (Fig. 3B). The resulting defect on the alveolar ridge was covered with a pedicled left buccal mucosa flap.

2.5. Microscopical findings

The mandibular soft tumor microscopically showed spindle to stellated cells disposed over a moderately loose matrix (Fig. 4A) and vimentin staining was positive (Fig. 4B). It was diagnosed as mesenchymatous proliferation.

The subjacent bone sample was composed of mature compact vital bone that changed into a pagetoid appearance with basophilic cementum lines resembling a mosaic, which was diagnosed as pagetoid bone (Fig. 4C).

It was suggested to further study the patient regarding the high level of alkaline phosphatase and the presence of pagetoid bone, as well as screening for additional lesions with the aid of a bone scintigraphy.

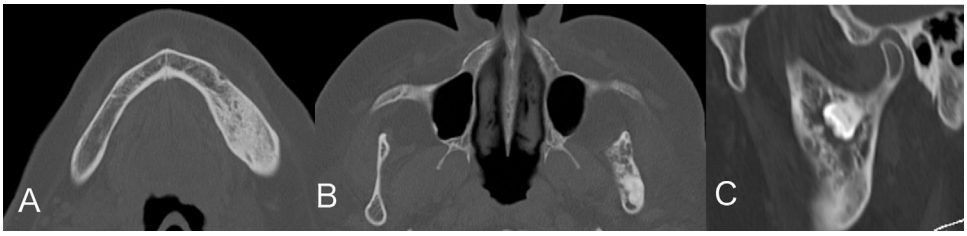

Fig. 2. A. Axial plane CT Scan exhibits ground glass appearance at the body of the mandible. B. Axial plane CT scan exhibits an heterogenoeous expansive process with cortical involvement at the mandible ramus. C. Sagital plane CT scan exhibits cranial and distal displacement of the left lower third molar.

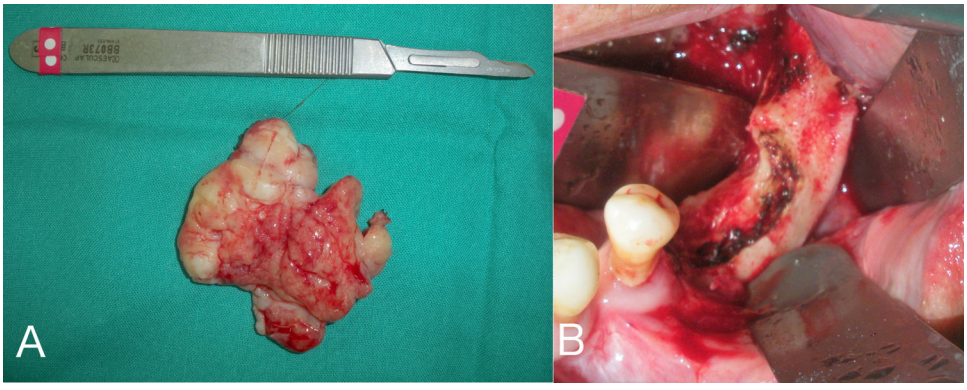

Fig. 3. A. Tumoral mass of the left cheek after complete excision. B. Alveolar ridge of the mandible after incisional biopsy sample was taken.

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