

Case Report Article

Surgical treatment of desmoplastic fibroma in the jaw using mandibular graft reconstruction – a case report

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Abstract

Introduction: Considered a rare benign tumour of fibroblastic origin, desmoplastic fibroma is an injury with aggressive behaviour and local recurrence and unclear aetiology, although trauma and genetic factors are considered. The pathology may develop in the femur, radio, pelvis and tibia, and rarely affects the jaws, characterized as an injury of slow asymptomatic growth that may vary radiographically. The local aggressive resection is the treatment of choice. **Objective:** To report a case of a desmoplastic fibroma in the mandible, whose treatment was *en bloc* resection followed by reconstruction with mandibular graft. **Case report:** Female patient, 33 years old, Caucasian, with a bone lesion discovered in routine x-ray. Images showed hypodense image with discrete destruction of bone cortical, near teeth 36 and 37, with no involvement of the roots. The chosen treatment was *en bloc* resection followed by reconstruction with mandibular graft harvest from ramus. **Conclusion:** desmoplastic fibroma is a benign lesion that should be included in the differential diagnosis list of osteolytic lesions. A long-term follow up is required, considering the potential to recurrence.

Introduction

The desmoplastic fibroma is an uncommon benign neoplasm of fibroblastic origin with an aggressive behaviour and local recurrence [10]. It is believed that the injury has no predilection for gender and may occur in patients younger than 30 years old, with a mean age of approximately 16 years old [12]. The World Health Organization defines desmoplastic fibroma as a rare tumour composed of spindle cells with mild atypia and abundant production of collagen [17]. The pathology may occur in any bone in the body, although femur, pelvis, tibia and radio are the most involved. When developed in the jaws, a rare condition reported only in 1965 [16], the main sites are angle and ramus [12].

Even though literature suggests trauma and genetic factors as possible causes for expression and development of the lesion, the exact aetiology remains unclear. Clinically, desmoplastic fibroma may present as an asymptomatic swelling that slowly grows [18]. Radiographically, may present different features, regarding the configuration of the lesion (unique or multilocular) and the margins (well or not defined) [12]. When left untreated, the lesion may cause bone destruction and tends to invade soft tissues. The rate of recurrence depends on the chosen treatment. The most recommended is *en bloc* resection or aggressive local excision [5]. Histopathologically analysing, one should consider fibrosarcoma, fibrous histiocytoma, fibrous dysplasia and even an intraosseous low-grade osteosarcoma as possible differential diagnosis [16].

Being so, this paper aims to report a case of a desmoplastic fibroma in the mandible, whose treatment was *en bloc* resection followed by reconstruction with ipsilateral graft of mandibular ramus.

Case report

Female patient, 33 years old, Caucasian, attended the Department of Oral and Maxillofacial Surgery of Erasto Gaertner Hospital, referring painful mandible lesion discovered in routine x-ray that developed after third molar extraction. The patient revealed central diabetes insipidus, hypercholesterolemia and hypothyroidism, treated with medication. Images showed radiolucent image, with about 2 cm diameter in the apical region of

teeth 36 and 37, of well-defined limits, without involvement of the roots and without apparent expansion of cortical bone (figure 1). CT scan showed hypodense lesion with a mild destruction of the cortical plate on the lingual side (figure 2). Incisional biopsy was performed in the region and, histologically, was consistent with benign spindle cell neoplasm. Among the clinical aspects, the main hypothesis was desmoplastic fibroma and so, the treatment proposed was aggressive *en bloc* resection and reconstruction of the defect with mandibular graft harvest from the ramus (figure 3). Postoperatively, the patient presented paraesthesia caused by resection of inferior alveolar nerve, since it was adjacent to the lesion and was sacrificed. The patient is in regular follow-up and, 4 months after the surgery, no complication or signs of recurrence have been observed (figure 4).



Figure 1 - Panoramic radiography evidencing a radiolucent lesion

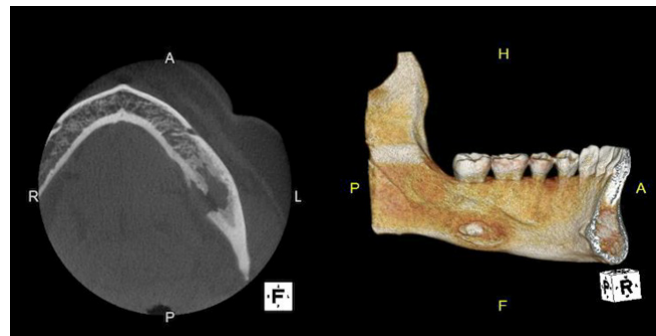


Figure 2 - Tomography in axial image of patient mandible showing a lesion in right body and 3d reconstruction

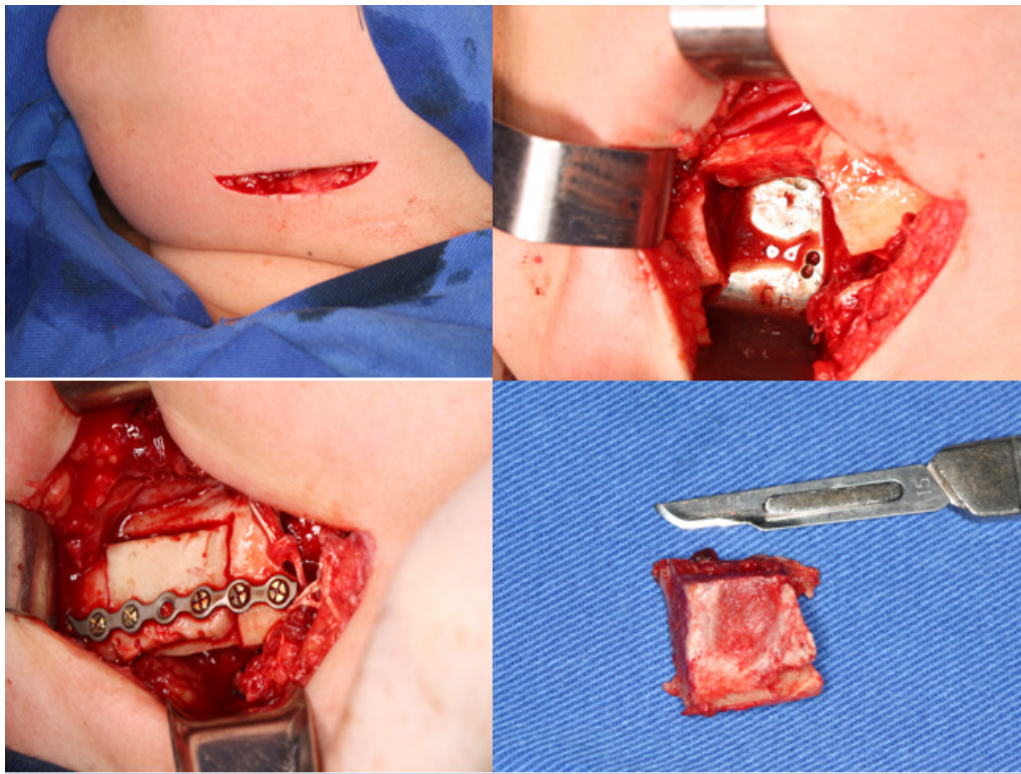


Figure 3 - Transoperative surgery showing resection of the lesion in jaw body with fixation of autogenous graft from the left mandibular branch

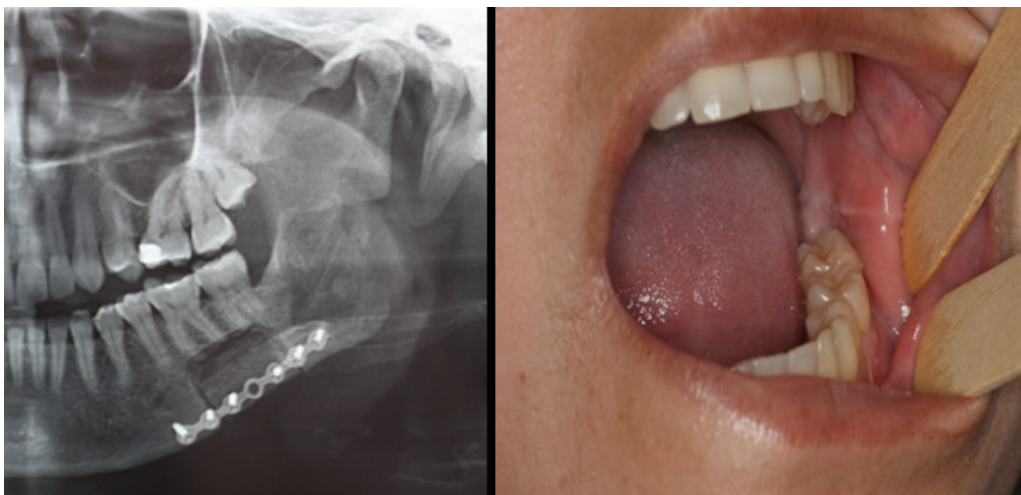


Figure 4 - Postoperative with 4 months of follow up

Discussion

The desmoplastic fibroma is a rare intraosseous tumour [8] first described in 1958 by Jaffe [7] and representing 0.1% of all benign bone tumours [15]. Although considered a benign lesion, it may be locally aggressive [7] and appear with high rate of recurrence if not properly treated [7].

A etiology remains unclear, even that a possible association with trauma, endocrine factors, genetic and multifactorial changes may exist [3]. The lesion may affect any bone in the body, but is commonly found in gnathic bones, femur, pelvic bones, radio and tibia [20]. In Averna *et al.* [1] reported less than 200 cases of DF described in the literature, less than 100 cases located in the jaw. When present

in the jaw bone, about 86% occur in the jaw and 14% in the jaw, with no apparent sex predilection [15, 16]. According to Freedman *et al.* [4], when located in the jaw, 70% of the lesion are in the molar area, 21% in the premolar region and only 9% in the anterior site.

The occurrence of desmoplastic fibroma has been reported in a wide age range, which can extend from birth to sixth decade of life [15]. However, it is more commonly diagnosed in patients under 30 years old [20]. Usually, there is no pathognomonic sign or symptom associated, although most of patients report pain and swelling [15]. Naief *et al.* [10] reported that 65% of patients had asymptomatic swelling, 15% complaint pain, 11% presented mouth-opening limitation, 7% tooth mobility, 2.6% proptosis and 2.6% presented local infection. In 19.4% of patients were not documented signs and / or symptoms. Radiographic features were reported by 91% of 74 cases. Thirty-six percent of patients showed a multilocular appearance while only 6% showed unilocular pattern. Only 4% of patients showed a mixed density [10].

The diagnosis of desmoplastic fibroma is difficult to achieve only by imaging studies. In both MRI and CT scan, many tumours resemble the image of this pathology, i.e. fibrous dysplasia, giant cell tumour, simple bone cyst, aneurysmal bone cyst, ameloblastoma and metastasis [11]. While CT evaluates the extent of bone destruction, MRI allows better visualization of the medullary portion as well as the extent of the tumour in soft tissue. Therefore, CT and MRI complement each other when investigating this lesion [19].

Histologically, desmoplastic fibroma is dominated by abundant collagen fibres and low density cells. The nuclei are elongated and fusiform and mitotic signals usually are not present [11]. The lesion is not encapsulated, which increases the difficulty on identifying the intraosseous limits [13]. The histological differential diagnosis is with low-grade fibrosarcoma, fibro-histiocytoma, fibrous dysplasia, low-grade osteosarcoma, aneurysmal bone cyst and juvenile bone cyst. Although fibrosarcoma presents a highly cellular stroma, along with high degrees of polymorphism and mitosis, low-grade variant may show a tissue rich in collagen with low cell counts and no mitotic activity [13]. Being so, a careful analysis should be performed in order to discard other possibilities [9].

Regarding the treatment, several modalities have been proposed, including surgery, radiotherapy and chemotherapy [10], although radiotherapy being not

recommended because of a slow success rate and high potential of development of radiation induced sarcoma [2, 10]. Among the surgeons, some prefer curettage, while others defend a wide local excision or resection with margin [10]. Shekhar *et al.* [18] reported recurrence of 20-40% of the cases treated with enucleation or local excision, while those treated only with curettage recurred in 70%. According to Patussi *et al.* [14], the patient underwent four operations in which all were performed in only curettage. When we opted for a more aggressive surgical approach and resection block in place the sprained success was obtained. It is noted that when an aggressive surgical technique is used, increases the success rate the prognosis of the patient [14]. The identification of the margins during surgery has been reported to be difficult because of the absence of a capsule delimiting the lesion. Currently, it has been recommended wide surgical margins of 2 to 3 cm where there is tumour infiltration in bone tissue with adjacent soft tissue invasion [6].

Although the literature usually does not report malignant transformation, Min *et al.* [9] reported the presence of secondary osteosarcoma in patients with a history of desmoplastic fibroma 10 years after before. However, malignant transformation in these cases was not confirmed by histopathology [9].

The case reported in this paper was consistent with the literature findings, since was in a young patient with an osteolytic lesion in the posterior region of the left mandible, region of molar teeth. The treatment proposed was *en bloc* resection, in order to give enough margin and reduce the possibility of recurrence, according to what is described in the literature. Despite its non-metastatic nature, the aggressiveness of the lesion and the high recurrence rates justify clinical and radiographic follow-up for a long period [18].

Conclusion

Desmoplastic fibroma is a benign tumour that should be included in the differential diagnosis of any osteolytic lesions, especially in young patients. The knowledge of the radiologic features and their correlation with histological findings is crucial for diagnosis and definition of the treatment, since the lesion has a lower tendency to recurrence when surgery is performed correctly, with a wide resection. It is also recommended a long term follow-up due this potential for recurrence.

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