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Case Report

Desmoplastic fibroma: Report of a rare case

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Abstract: Desmoplastic fibroma (DF) is a rare and locally aggressive benign neoplasm of connective tissue origin that is known as the intraosseous manifestation of soft tissue fibromatosis in both gnathic and extragnathic sites. The aim of this report is to define clinical, histopathological and radiographic features of a desmoplastic fibroma of the right mandible region in a 41-year-old male patient. The patient was initially presented with 6 month history of painless gingival swelling in the right mandibular molar area. Histopathological features and immunohistochemical evaluation allowed the differentiation of Desmoplastic fibroma from other lesions in the maxillofacial area.
Keywords: Bone neoplasms. Desmoplastic fibroma. Mandible. Jaw neoplasm.

INTRODUCTION

Desmoplastic fibroma is recognized as the osseous counterpart of aggressive soft tissue fibromatosis. In 1838 Johannes muller first characterized the term " desmoid" [1]. In 1958 Jaffe initially described a primarily osseous-arised aggressive fibromatosis of the femur, tibia and scapula [2]. He stated these tumors as " desmoplastic fibromas ". The first report about gnathic desmpoplastic fibroma was presented by Griffith und Irby in 1965 [3]. Since then several number of related cases have been published [4, 5]. Desmoplastic fibroma is a rare fibroblastic tumor, representing far less than 1% of all bone neoplasms and 0.3% of all benign tumors of bone. Mandible is reported as the most affected site in maxillofacial skeleton, however any bone may be involved. In extragnatic sites, it is most often seen in the metaphyseal region of long bones such as femur, tibia, humerus [4,6,7].

This tumor occurs equally in male and female patients and commonly affect patients younger than 30 years of age. Pathognomonic symptoms do not exist. Pain and swelling are occasionally reported. Incidental finding of lesion has also been noted. Desmoplastic fibroma often behaves in an aggressive manner .Radiologic features are nonspecific and the lesion could appear as a unilocular or multilocular radiolucency, with well-delineated or irregular borders. Aggressive pattern of bone destruction can sometimes be seen and the lesion may be misdiagnosed as malignancy [8,9].

As therapy, wide excision, resection, radiotherapy and if necessary pharmacological treatments have been used in managing this neoplasm. In respect of high recurrence rate and aggressive nature, surgical resection is the most recommended option [6].

The following is report of a case of desmoplastic fibroma in the right mandible region supplemented by its most important clinical and histopathological features.

CASE REPORT

A 41 -year-old male patient was referred to department of oral and maxillofacial surgery at the Taleghani Hospital of Shahid Beheshti University of Medical Sciences to be treated for a gingival swelling of 6 months duration at the right mandibular first molar region. The gingival enlargement was painless. Only buccal gingiva was affected by swelling. The patient had noticed the alteration 6 months before the intervention. First, he had visited a general dentist and

oral antibiotics was prescribed for his condition. With swelling persisting the patient was referred to us.

The patient had no history of trauma and past medical history was noncontributory. Clinical examination revealed an edentulous area between tooth #28 and #31 .The patient had his right second premolar and right first molar extracted two years ago due to progressive caries. The overlying mucosa was normal, there was no pulsation or tooth mobility. Extra oral examination showed no swelling. The remainder of the head and neck evaluation was within normal limits .The panoramic radiograph showed a unilocular radiolucent lesion with ill-defined limits measuring 2.5 × 1.5 cm extending from lower right first premolar to the lower right second molar. Displacement or resorption of adjacent teeth was not noted. (fig.1)

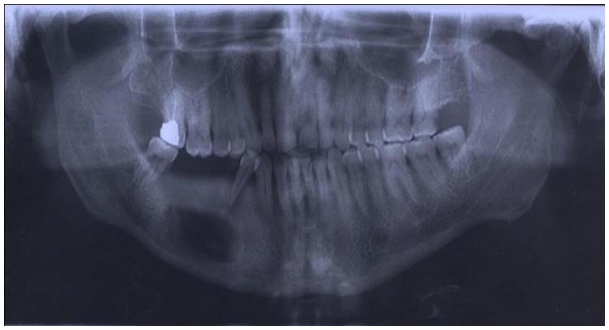

Fig-1: Panoramic radiograph reveals an ill-defined radiolucent lesion of the right mandible.

The patient underwent incisional biopsy under local anesthesia. The specimen was submitted for further histological evaluation .Microscopically, there was a foci of mature spindle-shaped cells proliferation which intermixed with abundant collagen fibers. The dense connective tissue exhibited low-cellularity and the extracellular matrix showed intense collagenization and was intensively hyalinizated. Neither pleomorphism nor mitotic figures were observed in any of the sections. (fig.2).

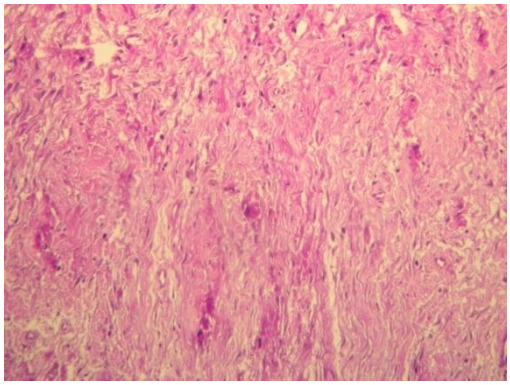

Fig-2: Microscopic section of tumor shows a mixture of thick bundles of collagenized fibrous tissue and aggregates of elongated spindle shaped cells. (Hematoxylin-eosin stain, original magnification ×100)

Paraffin-embedded tissue sections, fixed in formalin were stained with the following antibodies according to manufacturer recommendations: anti S-100 protein and anti-vimentin. The tumor cells were nonreactive for S-100 protein but showed strong and diffuse immunoreactivity for anti-vimentin (fig. 3-6)

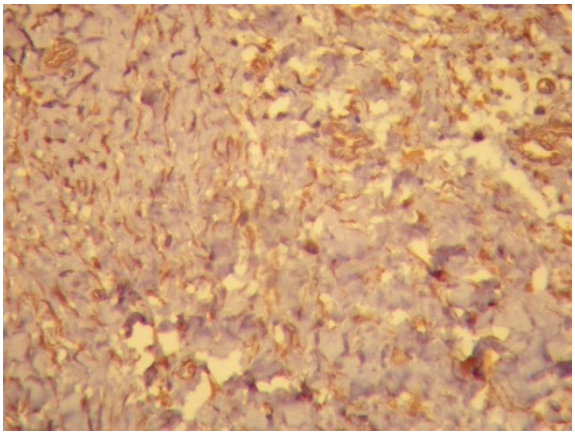

Fig-3: The tumor cells showed strong and diffuse reactivity for anti-vimentin (original magnification ×200).

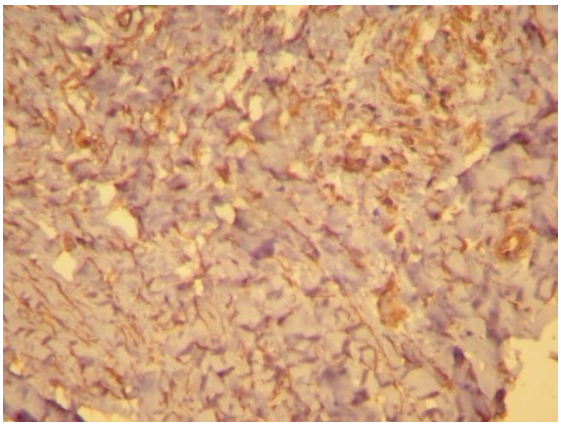

Fig-4: The tumor cells showed strong and diffuse reactivity for anti-vimentin (original magnification ×400).

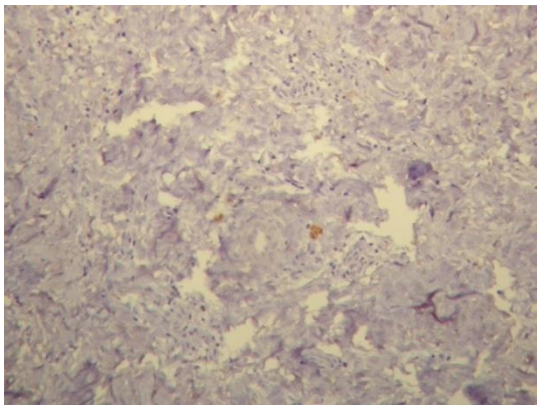

Fig-5: The tumor cells were nonreactive for S100 protein (original magnification ×100).

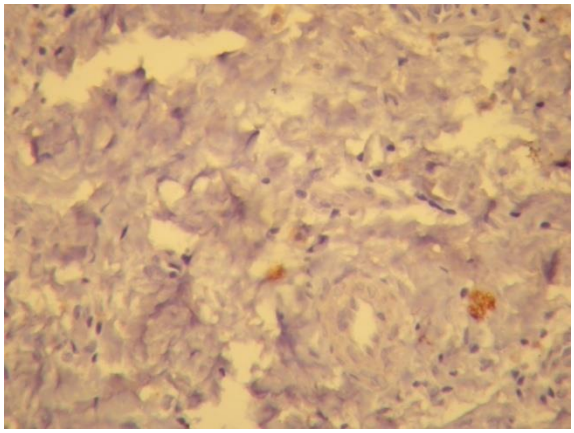

Fig-6: The tumor cells were nonreactive for S100 protein (Original magnification ×400).

Based on the clinical, radiological, and pathological appearance, a final diagnosis of Desmoplastic fibroma was rendered.

Subsequently, surgical removal of the lesion under general anesthesia was undertaken intraorally. Using a sulcular incision from tooth #27 to #31, wide excision of the lesion was performed. The tumor was not adherent to the surrounding tissues. Gross examination of the excised specimen revealed creamy-brown elastic to firm, non-capsulated tissue measuring 1.5×1×1 cm. Tooth #28 was extracted during the surgical operation. The patient tolerated the procedure well and was discharged the day after the surgery .Following the removal of the lesion, swelling was completely resolved after 7 days. The patient remains under review .The time for the scheduled follow-up appointments has not arrived yet.

DISCUSSION

The World Health Organization describes the histological criteria for desmoplastic fibroma as that of a benign bone tumor of low to variable cellularity and abundant collagen production. The tumor cells can be ovoid or elongated with uniform nuclei and no increase in mitotic activity that lack atypia and pleomorphism. A matrix of collagenized fibrous connective tissue with diffuse hyalinization support the tumor cells. Histologically, it mimics the desmoid tumor of soft tissues [10].

The etiology is unidentified, although trauma and endocrine and genetic factors have been proposed. Trauma to the affected area is the most possible precipitating factor [5,11]. The tumor is more frequent during the second and thirds decades of life. Although incidence in the maxilla has been described, the mandible is the most common facial region, with most lesions in the ramus-angle site. Generally, patients are 15.1 years old at the time of the final diagnosis and there is no apparent sex predilection. Our patient was a 41 years old man with a lesion at the posterior right mandible region. [12,13]. In long bones, a usual presenting sign of the disease is pathologic fracture. In the facial skeleton, however, a gradual swelling, asymmetry, or both most commonly alert the patient or clinician to its presence. Occasionally pain and functional impairment exist. Our patient had the chief complaint of painless gingival swelling at the right mandibular first molar site.

In general, the radiographic features of DF are nonspecific. These include a unilocular or multilocular, well-demarcated or irregular radiolucency. Marginal sclerosis and cortical rupture are less common [14]. Therefore ,radiographically DF often mimics other common as well as unusual pathologies of the jaws including ameloblastoma, odontogenic myxoma, aneurysmal bone cyst, chondromyxoid fibroma, central hemangioma , and eosinophilic granuloma [15]. In our case, ill-defined unilocular radiolucency was noticed. The orthopantogram did not show any sign of cortical disruption. The radiographic features of the case presented are compatible with the patterns described.

The rapid growth and bone destruction often seen in association with DF, may wrongly lead to an inaccurate diagnosis of malignancy .Owing to the high recurrence rate and aggressive behavior of the lesion, the differentiation of DF from other bone lesions in the maxillofacial skeleton is crucial, bearing in mind it has a significant impact on the surgical management of the lesion [16].

Microscopically, the tumor is characterized by relatively monotonous appearance with an absence of mitotic figures .The extensive areas of collagenization, diffuse hyalinization and spindle-shape fibroblasts should be emphasized. All of these findings were present in our case. Another important aids in DF differentiation are immunohisthochemical aids, as DF is immunopositive for vimentin [16,17]. Since in some parts of the microscopic appearance tumor cells were spindle with wavy nuclei, similar to neural cells, S100 protein was used to rule out neural tumors.

Histologically, it is sometimes challenging to distinguish desmoplastic fibroma from other fibrous lesions. One of the most difficult differential diagnosis from DF is low-grade, well-differntiated fibrosarcoma. Important distinguishing features of fibrosarcoma are increased mitotic activity, extensive nuclear pleomorphism, a greater density of cells and paucity of collagenous background. Unlike fibrosarcoma where cells usually assume fascicular growth pattern and often produce a so called "herring bone" appearance, DF favors a single cell orientation and may be seen arranged in bundles. In this case, no evidence of malignancy was seen [18].

In the present case, the final diagnosis of DF was only possible through histopathological examination and immunohistochemistry

staining.Variable treatment modalities have been used for DF including surgery, radiation therapy, and chemotherapy with or without additional procedures. Radiation is not recommended as a primary treatment since it has been shown to be only rarely effective and, because of its potentially mutagenic effect, may lead to postradiation sarcoma. In some situations when the surgical operation is not possible or the patient objects to the surgical procedure, the radiotherapy can be used as an alternative. Varying surgical procedures have been used in managing the desmoplastic fibroma, including local curettage, wide excision, resection, or mandibulectomy. In our case wide local excision with curettage was the treatment of choice. The prognosis for patients with desmoplastic fibroma is dependent on the completeness of surgical removal. According to the literature, patients treated with resection or wide excision showed no recurrence. The infrequent use of enucleation is due to the concern that recurrences rates are higher, approximately 20-40% compared to a minimal recurrence rate in block resection or wide local excision [6]

Kwon et al reported that tumors with high cellularity have higher recurrences than those with lower cellularity. In our case, microscopic evaluation of the lesion showed low cellularity. Because of the tendency for local recurrence, the follow-up period should not be less than 3 years [19]. The time ime for first session of follow-up has not arrived yet for our patient.

CONCLUSION

This case illustrates the need to include rare pathological conditions in the initial diagnosis of lesions in the maxillofacial area .Correlation of clinical, radiologic , and histologic findings is imperative for determining a definitive diagnosis, whilst always being aware that the cause of a gingival swelling may not be as simple as it often seems.

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