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Case report

Desmoplastic fibroma of the mandible associated with pathological fracture in a pediatric patient

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ABSTRACT

Desmoplastic fibroma (DF) is a benign, locally aggressive, bone tumor that rarely presents in the facial bones. It is considered an intraosseous counterpart of the soft tissue fibromatoses. It is very rare in children. The present report describes the clinico-pathological and radiographic features of a desmoplastic fibroma of the mandible in a 12-year-old female who presented with a few months history of an expanding, painless mass along the left inferior border of the mandible. The patient underwent a resection with wide margins of the inferior border of the mandible that was followed by a pathological fracture. The fracture was reduced and the mandible was reconstructed by a vascularized free fibula flap.

Conservative surgical approach and long term follow up is an effective treatment modality for mandibular DF in children.

1. Introduction

Desmoplastic fibroma (DF) is a rare, benign, locally aggressive tumor of bone. Any bone can be affected, but the mandible is the most frequently affected site. It is recognized as the osseous manifestation of the soft tissue fibromatosis. The involvement of multiple bone sites, such as the femur, pelvic bones, radius, and tibia have been reported [1–4]. The first report on mandibular involvement is attributed to Griffith and Irby in 1965, and since then numerous cases have been described [5–7].

DFs occur most frequently in young adults and present as a slow-growing, painless swelling. It is considered rare in children.

The surgical approach to DF is controversial. Variable treatment modalities, ranging from conservative enucleation to a complete en bloc resection, with different outcomes, have been implemented [6]. Radiotherapy or chemotherapy is indicated, for patients with non-resectable tumors [4].

The purpose of this article is to report on a case of DF occurring in the mandible of a 12-year-old female patient, its diagnostic work-up, initial conservative surgical management, complication by a pathological fracture and the reconstruction by a vascularized free fibula flap.

2. Case report

A 12-year-old female presented with a swelling on her face which was first noticed two months previously. Extra-oral examination revealed a mild facial asymmetry and swelling of the left mandible. Oral examination showed some tenderness in the floor of the mouth adjacent to the left mandible. All the permanent teeth were intact and stable. The remainder of the orofacial examination was within normal limits. The patient's medical history was noncontributory. However, her mother had many abortions and died during pregnancy for an unknown reason. There is no other family history of cancer or skin disease.

A panoramic radiograph revealed a unilocular radiolucent lesion with disruption of the left inferior cortex of the mandible. The lesion extended from the inferior border in the premolar-molar region (Fig. 1A). Axial CT of the mandible demonstrated a lesion at the posterior mandible that is extending toward the soft tissue adjacent to the lingual side of the mandible (Fig. 1B).

Under general anesthesia, a biopsy from the center of the lesion was obtained and submitted for histological evaluation.

Microscopic examination revealed an ill defined spindle cell proliferation in a collagenous background with varying degrees of myxoid changes. The arrangement of the spindle cells varied with

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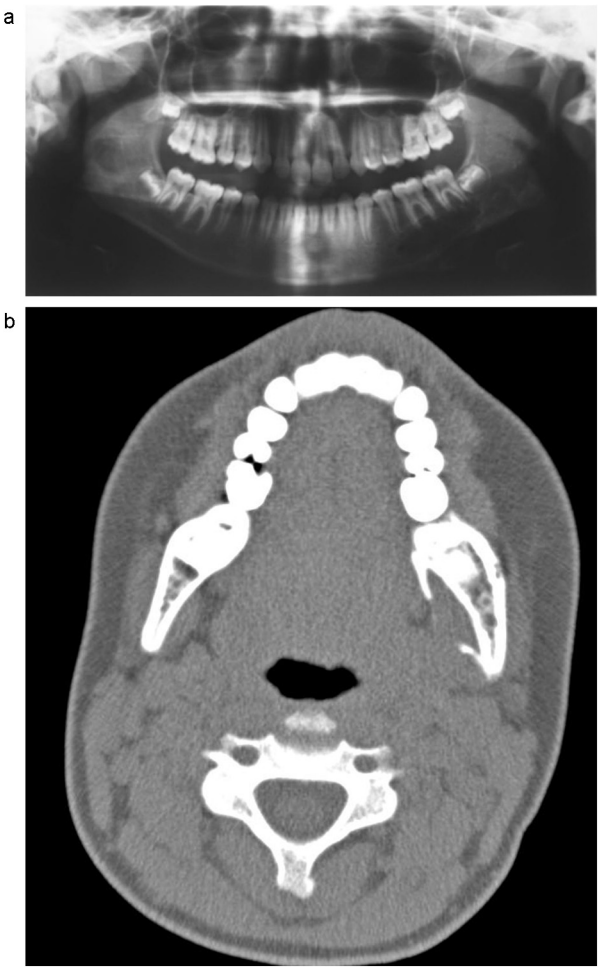

Fig. 1. (a) Preoperative panoramic radiograph depicting a unilocular radiolucent lesion at the left body of the mandible. The lesion extends from the inferior border in the premolar-molar region. (b) Axial CT of the mandible showing lesion expanding from the left mandible and medially into the soft tissue as can be seen by loss of fat planes.

groups of cells arranged in parallel fashion mixed with short interlacing fascicles and intertwining strands. The spindle cells were generally uniform in appearance. Nuclear atypia, mitotic figures or necrosis were not present. The tumor was seen in bone and soft tissue (Fig. 2a).

Immunohistochemically the tumor cells showed focal reactivity for smooth muscle actin and strong and diffuse reactivity for vimentin. Tumor cells were nonreactive for S-100 protein and muscle-specific actin. The Ki67 index was low (1–3%) (Fig. 2b).

Based on the clinical, radiological and histopathological appearance, a diagnosis of DF was made.

The patient underwent a resection with wide margins of the left inferior border of the mandible, via a submandibular approach. No grafting was performed. All the molars of the left mandible were extracted to reduce the masticatory forces. Final pathology report was consistent with desmoplastic fibroma extending into the submandibular gland (Fig. 2).

The short term postoperative course was uneventful. At 6-month follow-up visit the patient complained of some discomfort during mastication. Clinically, some mobility at the left body of the mandible was noticed. Panoramic radiograph, demonstrated a pathological fracture of the body of mandible with severe dislocation, distal to the second premolar (Fig. 3). MRI study of the left submandibular area, depicted no recurrence at or adjacent to the fracture site. Open reduction and internal fixation (ORIF) via a submandibular approach, was undertaken, using a 2.0 mm craniomaxillofacial fixation system (W. Lorenz Surgical, Jacksonville, FL, USA). 2.0 mm, straight, regular, monocortical mini plates were used for both reduction of the fracture line and for stabilization of the vascularized free fibula flap.

The initial post operative course was unremarkable, however, at four weeks after surgery the patient presented with an abscess in her left submandibular region. The abscess was drained percutaneously followed by frequent irrigation and antibiotic therapy with complete recovery. Five months later bone healing can be seen (Fig. 4).

The patient has been followed for the past 26 months without any clinical or radiographic signs of recurrence.

3. Discussion

Fibromatosis of the head and neck area, may be intra osseous (such as DF) or more often, in the soft tissue, mainly in the supraclavicular region of the neck [17].

DF is defined by the World Health Organization (WHO) as a benign tumor of low to variable cellularity, whose cells can be ovoid or elongated with uniform nuclei that lack atypia, pleomorphism and mitotic activity [8]. Though similar in name it should be differentiated from desmoplastic fibroblastoma [9] and desmoid tumors [10].

The etiology of DF is still unclear, however, trauma, endocrine, and genetic factors have been suggested as possible etiologic factors [19]. In our present case no trauma or endocrine factors can be a possible cause. Genetic factor, obviously can never be ruled out. As no cytogenetic analysis was done in the present case we cannot comment on it.

DF may be involved with any bone, but the mandible is the most common site of involvement (22%), followed by the femur (15%), pelvic bones (13%), radius (12%), and tibia (9%) [2].

The conditions commonly responsible for pathological fractures of the mandible, include osteoporosis, osteomalacia, Paget's disease, osteitis, osteogenesis imperfecta, benign and malignant bone tumors. These factors, however, are more common among elderly patients. Pathological fracture of the mandible is generally uncommon and it is considered rare among children [11,12].

Two previous reports exist of a pathological fracture of the mandible caused by DF, however both were in elderly patients [13,14]. Our present case is the first report in a pediatric patient. We do not know whether the fracture occurred due to the disease, as a result of the surgery or both. Obviously both can cause the remaining mandible to be too weak to withstand the stress of mastication.

Based on MRI study, no recurrence was exhibited at or adjacent to the fracture site, indicating that the fracture was not associated with recurrence.

Desmoplastic fibroma was named in the past as desmoid of the bone. This is because of the histological similarity between DF and desmoid tumors which originate from soft tissue. In many cases one cannot tell whether the tumor began from soft tissue invading the bone or vice versa [15]. In the present case the tumor clearly originated in the mandible and extended into the soft tissue of the submandibular area.

Low-grade fibrosarcoma is the most challenging differential diagnosis and the most important lesion to differentiate from DF. In fibrosarcoma, the cells have a fascicular growth pattern and often produce a so-called "herring bone" appearance, whereas DF favors a single cell orientation and may be seen arranged in bundles. Also, overlapping of the spindle cells, increased mitotic activity, pleomorphism, and paucity of the collagenous background are characteristic features of fibrosarcoma and not DF. In certain circumstances, the distinction between the two conditions,