

DESMOPLASTIC FIBROMA WITH PERIOSTEAL REACTION

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Abstract- Desmoplastic fibroma is a rare, locally aggressive, benign lesion. A case of desmoplastic fibroma of left side of mandible is reported. The lesion presented as painless mass in left posterior portion of mandible of a 7-year-old girl. Histologically the lesion was composed of interlacing fascicles of benign appearing fibroblast in a varying ground substance of collagenous tissue. In this report, we discuss on radiographic findings and compare them with previous reports.

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INTRODUCTION

Desmoplastic Fibroma was described in 1958 by Jaffe. This Tumor has been described as a rare, locally aggressive, benign lesion. Desmoplastic Fibroma (DF) is considered to be the intraosseous counterpart of the soft tissue fibromatosis or desmoid tumor (1). This tumor is frequently found in the long bones, the mandible and the pelvis (2, 3) but ten cases of DF within the cranium have been reported (4). High incidence of this lesion is before the age of 10 years irrespective of significant sex predilection (5). The initial symptoms include swelling of the jaw and occasionally loss of teeth (6). Radiographic features of this lesion are usually expansive multilocular radiolucency with ill-defined border and very coarse and thick internal septa (7).

Microscopically, fibromatosis is characterized by infiltrating mature spindle shaped fibroblast arranged in streaming and interlacing fascicles with prominent mature collagen production, but cellular mitoses or atypia is not present (8).

Histopathologically, it is very difficult to distinguish desmoplastic fibroma from grade I fibrosarcoma and grade I fibroblastic osteosarcoma (9).

CASE REPORT

A 7-year-old girl presented with left submandibular swelling two months ago. She did not complain from spontaneous pain. A history of trauma was not found. On clinical examination, there was a firm swelling on left submandibular area with extension on buccal surface of posterior portion of mandible. Tenderness by palpation of swelling was not detected. Overlying skin on above area was normal. She was able to open her mouth without any limitation. Clinically, pinprick sensation deficiency on left lower lip and gingival portion was not detected. A panoramic view revealed that there were multiple radiolucencies with permeative pattern extending from the left mandibular first molar to left angle of mandible. These lucencies were distributed among upper and lower areas of mandibular canal. Apparently, mandibular canal was not involved. Enlarged follicle of left second molar tooth with lateralization of calcified follicle was also defined (Fig. 1).

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Desmoplastic fibroma

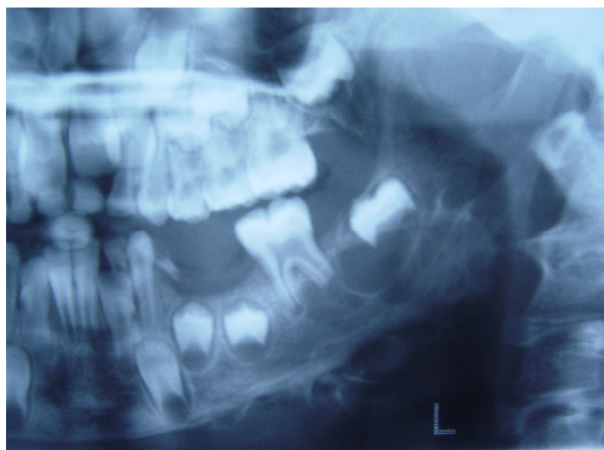


Fig. 1. Panoramic view made on 30 June 2004 showing multiple radiolucencies with permeative pattern and irregular radiated bony spicules from inferior border of mandible in LT side.

Radiating irregular bony spicules as a periosteal reaction arising from inferior border of mandible was observed. Lateral cross-sectional occlusal view and PA mandible view revealed expansion and perforation of lingual cortex. Fine and loose trabeculation was also detected as internal structure.

Early differential diagnosis was Ewing's sarcoma, osteosarcoma, lymphoma and histiocytosis X respectively.

After incisional biopsy from lesion, histopathological examination demonstrated a cellular lesion composed of benign appearing spindle-shaped cells arranged in interlacing fascicles and variable amounts of collagen fibers. Some spicules of adjacent bone along with reactive bone formation were also seen (Fig. 2). Final microscopic finding was "Desmoplastic Fibroma" of mandible.

Segmental resection was suggested by maxillofacial surgeon as the treatment of choice. CT scan was also ordered for the patient. But patient's parents did not accept this treatment. Finally, patient was operated and the lesion was curetted by general surgeon on 16 September 2004. Result of histopathology finding was confirmed Desmoplastic Fibroma. Patient was again referred us on 5 February 2005 by a dentist due to swelling in left side of mandible, mobility of left mandibular first molar and tissue bulging in this area. New panoramic view showed floating tooth appearance of left mandibular first molar with root resorption.

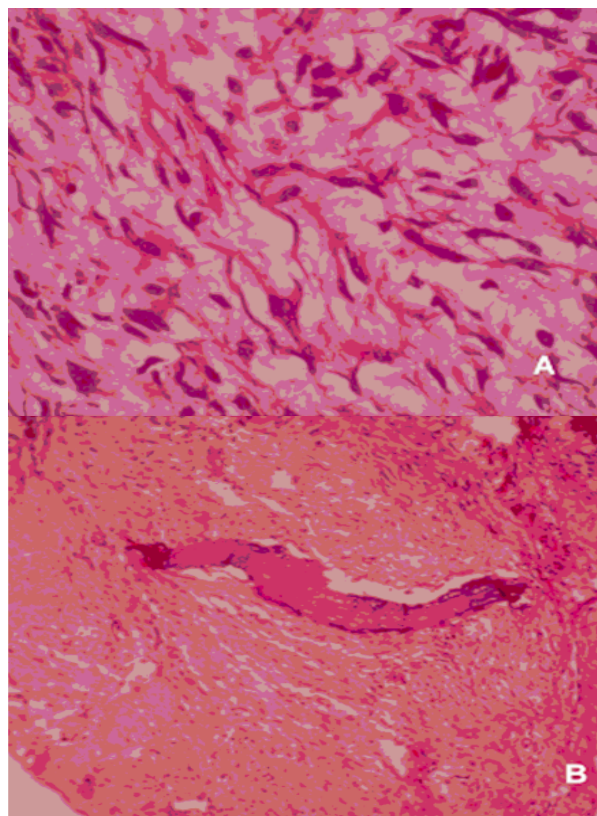


Fig. 2. Hematoxylin and eosin-stained section showing (A) interlacing Fascicles of spindle-shaped cells with intervening collagen (original magnification $\times 400$), (B) Tumoral cells with bony spicules (original magnification $\times 100$).

Remaining regular bony spicules arising from inferior border of mandible had been observed. There was no healing sclerotic border around the surgical site of area above mandibular canal.

Discontinuity of upper cortical border of mandibular canal was seen in one point (Fig.3). Spiral 3D CT Scan was provided. Axial CT slices through the body of mandible at the level of mandibular alveolar crest and at the level of inferior border of mandible, and also coronal CT in the level of posterior portion of mandibular body demonstrated significant expansion of lingual cortex with pressure effect on genioglossus muscle and lateral wall of oropharynx. Bony spicules arising from inferior border of mandible was also detected (Fig.4). 3D spiral CT image indicated extension of bone defect on inferior border of left side of mandible in comparison with right side (Fig. 5). Conclusively, lesion recurred. Patient's father did not agree with surgical treatment.

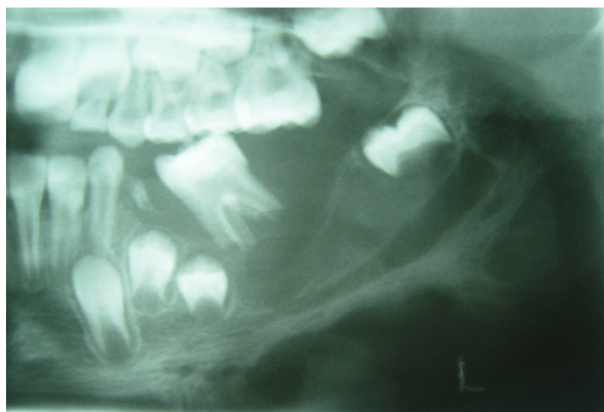


Fig. 3. Panoramic view on 5 February 2005 reveals remaining irregular bony spicules, floating appearance of left mandibular first molar and root resorption.

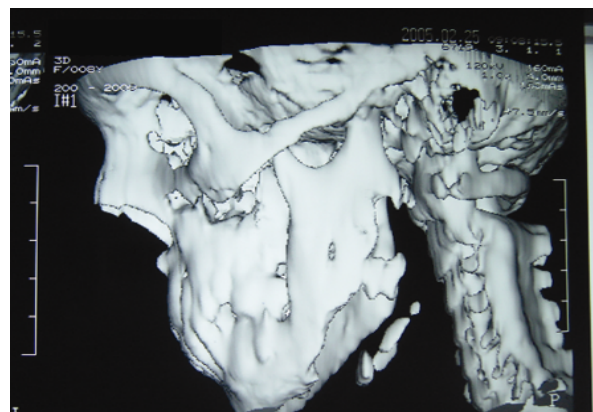


Fig. 5. Three dimensional reconstruction showing extension of bony destruction of inferior border of mandible.

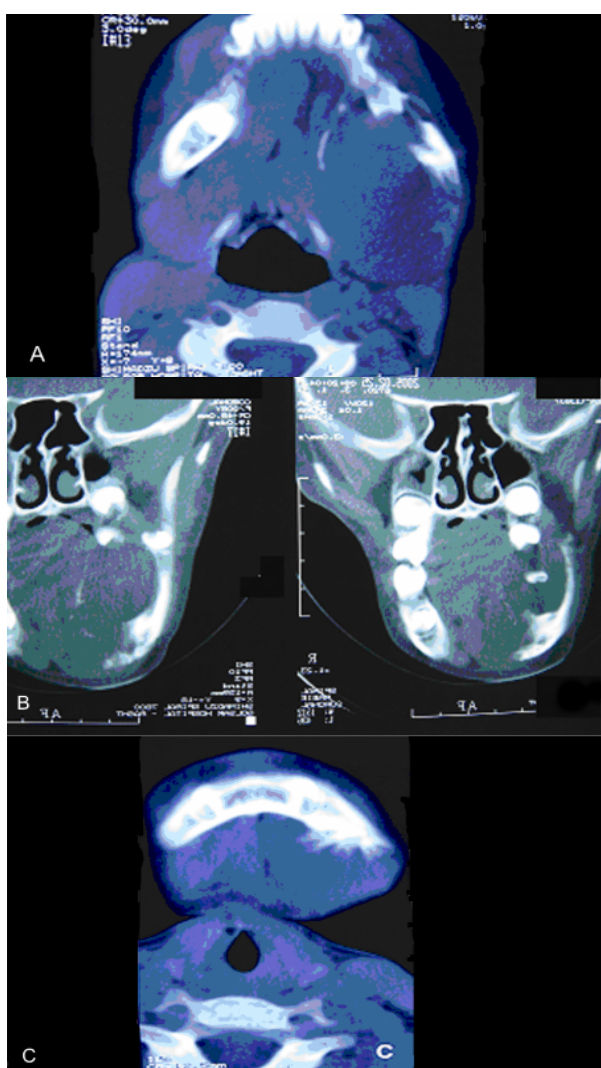


Fig. 4. Axial CT scan, (A) bone window; at the level of alveolar crest, and (B) axial T-bone window; at the level of inferior border of mandible, (C) coronal CT, bone window; at the level of posterior portion of mandible demonstrating expansive mass with significant thinning of lingual cortex, irregular spicules near inferior border of mandible.

DISCUSSION

Desmoplastic Fibroma is an aggressive and infiltrative neoplasm that produces abundant collagen fibers. The most common complaints of patient with desmoplastic fibroma are facial swelling and sometimes dysfunction. Pain is reported in rare cases. The age incidence is in the first two decades of life with a mean reported age of 14 years. It also may occur as part of Gardner's syndrome (7).

Most of the mandibular lesions are posterior to the molar region in the angle-ramus area (7). The most common reported radiological features of Desmoplastic Fibroma were geographic pattern of bone destruction with narrow transition zone and non-sclerotic margins, internal pseudo trabeculation (coarse thick septa or thin lacelike pattern) and cortical expansion (10). Perforation of the cortex was also present. New periosteal bone formation was seen in a limited number of reported patients (7, 10, 11).

Hashimoto and colleagues (1991) analyzed the radiological findings in 47 cases of desmoplastic fibroma; well defined multilocular radiolucency was common radiographic finding in these series. Poorly circumscribed radiolucencies were described in twelve mandibular cases (11). Irregular radiolucency was also reported for this lesion (6, 10, 11)

CT or MRI is suggested for detection of the exact soft tissue extension of the lesion. The radiographic appearance may not be effective in differentiation of this lesion from malignant lesion such as fibrosarcoma, malignant histiocytoma and

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osteosarcoma. Histopathologically, fibrous dysplasia and low grade osteosarcoma can be differentiated from Desmoplastic Fibroma by recognising areas of bone formation and nuclei form (3).

Low grade fibrosarcoma (9, 3) and fibroblast predominant osteosarcoma (9) are the most difficult problem in the histological differential diagnosis.

These tumors, although benign, are locally aggressive and can recur with a subtotal resection. For patients with Desmoplastic Fibroma arising from maxilla or mandible with extraosseous extensions, complete excision including a margin of uninvolved soft tissue is recommended (12, 13).

Lesion curettage or marginal excision develops a recurrence. This event has also happened in our case within 5 months after treatment. This finding confirms that Desmoplastic Fibroma with the reactive bony rim requires wide excision for prevention of recurrence.

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