

Desmoplastic Fibroma of the Maxilla : Report of a Rare Case

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Abstract:

Desmoplastic fibroma (DF) is a rare, benign but locally aggressive, intra-osseous lesion with a high tendency of local recurrence. A rare case of desmoplastic fibroma in the maxillary corpus is being presented with its pathology, clinical diagnostic methods, treatment and prognosis.

Key Words: Desmoplastic, Fibroma, Maxilla.

Introduction:

Desmoplastic fibroma is a benign, locally aggressive, intra-osseous tumor that rarely involves the facial bones. It represents the intra-osseous counterpart of the soft tissue fibromatosis or desmoid tumor. Jaffe & Selin in 1951, were the first to use the term desmoplastic fibroma to describe the lesion as a separate fibrous tumor of the bone. The first report about a desmoplastic fibroma of the jaw was presented by Griffith & Irby in 1965. In jaw area, non-odontogenic fibromatosis was declared as desmoplastic fibroma which distinguished it from Odontogenic fibroma (Slootweg & Muller, 1983; Depprich et al, 2005).

It is an extremely rare tumor with less than 200 cases in the published literature with a reported incidence of 0.11% to 0.13% among primary bone tumors (Bohm et al, 1996). It occurs more often in the first 3 decades of life with equal occurrence in men and women (Crim et al, 1989).

As therapy, surgical resection, radiotherapy and if necessary, pharmacological treatment are recommended. Due to high recurrence rate, surgical resection is the most preferred option (Sinno & Zadeh, 2009; Ikeshima & Utsunomiya, 2005).

In this report we present the clinical course and treatment of a patient with the diagnosis of desmoplastic fibroma in the right maxillary corpus, which itself make it rarest of the rare occurrence.

Case Report:

A 17-year-old male patient reported to the Department of oral and Maxillofacial Surgery with the complaint of swelling over his right cheek causing facial

asymmetry. Patient gave the history of swelling being a pea sized growth since childhood which gradually increased in size with age. The swelling was not associated with any pain or discharge, and the patient was mainly concerned with the distortion of the face caused by the swelling. On inspection, a solitary oval swelling with diffused margins of approximately 5 X 6 cm was seen extending from the right angle of mouth to 1cm lateral to right angle of mandible mesio-distally, and extending from 1cm below the right zygomatic buttress till lower border of mandible. On palpation, the swelling was found to be firm in consistency, lobulated, non-tender and fixed to underlying structure with no rise in local temperature.

Intra oral examination revealed an exophytic lobulated oval mass measuring 3 X 2.5 cm and hanging from buccal alveolar mucosal aspect in relation to 15, 16 & 17 (Fig. I). Vestibular obliteration was present with respect to right buccal mucosa. On palpation, it



Fig.I: Exophytic lobulated mass seen over right buccal mucosa.

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was lobulated and firm in consistency. Superiolateral aspect in relation to buccal vestibular area of 16, 17 had hard bony mass fixed to underlying structure, displacing 15, 16, 17 palatally.

Orthopantomogram did not reveal any gross bony change. Computerized tomography scan revealed, a fibro osseous growth over right maxilla, just posteriolateral to molar teeth causing subcutaneous elevation of soft tissue and the lesion was centrally located (Fig. II). Three Dimensional CT reconstruction revealed expansion of right maxilla anteriorly.

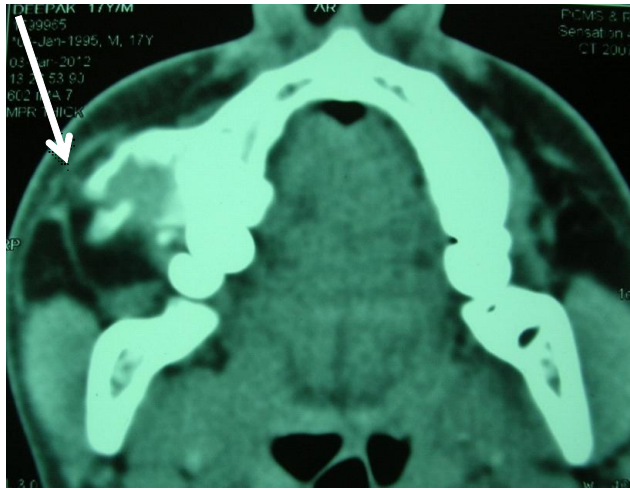


Fig.II: CT scan revealed fibro osseous growth over right maxilla.

Fine needle aspiration cytology was negative and therefore, incisional biopsy was done which was suggestive of desmoplastic fibroma. Routine blood and biochemical examination did not reveal any significant changes. Surgical resection of the tumor and curettage of the peripheral bone was planned under GA.

The tumour was excised and 17,18 were extracted because of its direct association with the tumour mass. Maxillary bony expansion was reduced using vulcanite burr and bone files and primary closure was achieved.

This tumour showed a very slight adherence to the surrounding bone and was resected easily. The histopathological examination of the resected mass revealed spindle-shaped cells with myofibroblastic differentiation, abundant collagen formation and low proliferation activity (Fig. III).

Based on these characteristics, the diagnosis of a desmoplastic fibroma was made. The post-operative recovery was normal. One year follow up of the patient showed no recurrence (Fig. IV).

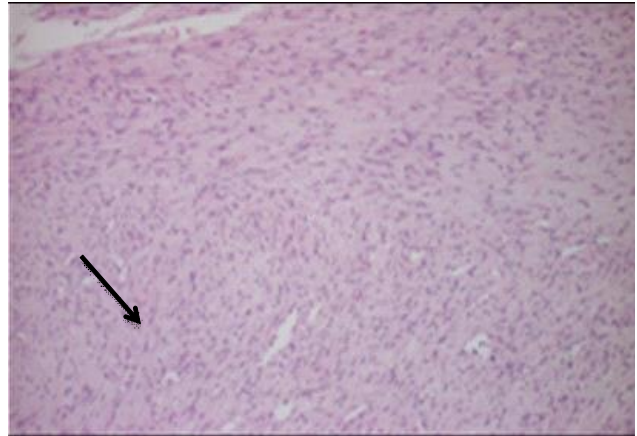


Fig.III: Partially parallel or plexiform arranged spindle cells with slim and elongated nucleus without cytological signs of malignancy. High-grade formation of collagen fibre. (H & E Stain, 100X).



Fig.IV: Patient after 1 year follow up.

Discussion:

Desmoplastic fibroma of the maxillofacial region may be difficult to diagnose early in the disease process because of a slow, insidious onset coupled with unremarkable radiographic findings. It is a disease of young people, with a peak incidence in the second decade of life (Templeton et al, 1997).

Rabhan & Rosai (1968) and Kwon et al (1989) reported that a desmoplastic fibromas of the jaw with increased cellularity have a higher tendency to recur. Controversy exists regarding the preferred method of surgical management. Jaffe & Selin (1951) recommended segmental resection, but also stated that thorough curettage was an acceptable alternative.

Freedman et al (1978) found that 19 of 22 cases of desmoplastic fibroma had no evidence of recurrence, with follow-up time ranging from 3 months

to 8 years. He concluded that curettage was the preferred treatment.

In the present case, excision of the tumour mass with curettage of surrounding bone was the preferred treatment because of non-aggressive nature of the lesion demonstrated by slow progression of the swelling over a duration of 17 years and the elevation of soft tissue rather than extension radiographically. Isolated intra-osseous lesion without evidence of extension into contiguous soft tissue or perforation of cortical bone may be adequately managed by thorough curettage. However, when a lesion displays signs of aggressive behaviour and extension into soft tissue, segmental resection should be considered.

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