CASE REPORT

Odontogenic Fibromyxoma

S Gupta, R Gupta, A John, H Umarji

Abstract

Odontogenic Fibromyxoma is a rare mesodermal tumour found exclusively in the bones of the facial skeleton. Although benign, it is locally very aggressive making it difficult to eradicate. We are reporting a case of odontogenic fibromyxoma in the maxilla with review of literature.

Key Words

Odontogenic fibromyxoma, Mesodermal tumor, Maxilla

Introduction

Odontogenic fibromyxoma is a rare odontogenic tumor of mesenchymal origin. It is composed of large amounts of intercellular substance rich in acid mucopolysaccharides making it locally very aggressive and with high recurrence rates following consevative excision. Its histological and radiological features make it difficult to differentiate from other odontogenic tumors and occasionally may be misinterpreted as a malignant lesion. In this paper, a rare case of odontogenic fibromyxoma of the maxilla in a 27 years-old women is presented with emphasis on review of relevant literature, histological and radiological aspects in differential diagnosis, its malignant potential and management.

Case Report

A 27-year-old woman presented with a history of a swelling of 7-month duration in the right maxillary alveolus. She gave a history of a swelling in the right maxillary canine area which had been painful, mobile, exuded pus and episodes of fever. She was prescribed antibiotics and the tooth had been extracted 4 months ago. The pain had subsided and she had been afebrile since then although the swelling had still persisted and had gradually increased in size. She now complained of dull, diffuse and chronic pain on right side of the face, which increased on opening the mouth. She also complained of heaviness on the right side of the face. There was no relevant medical history besides chronic maxillary sinusitis and she did not have any oral habits.

On examination there was a diffuse tender swelling in the right canine fossa region causing obliteration of the right nasolabial fold. Intraorally there was expansion of both the cortical plates in the right maxillary alveolus associated with a missing canine (Fig.1). The swelling was bony hard to palpation with normal overlying mucosa. There was no regional parasthesia /anaesthesia.

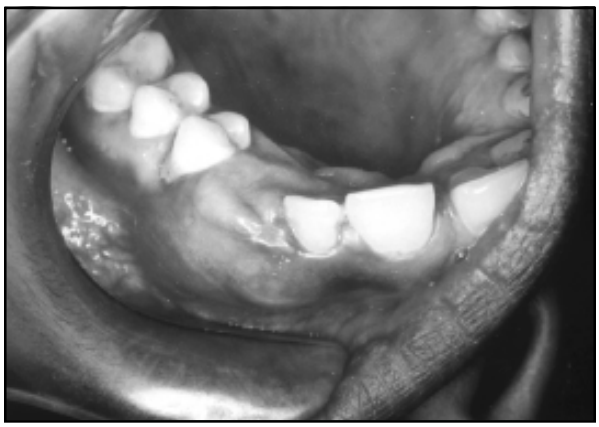

Fig- 1

From the Department of Medicine, AIIMS, Oral Medicine Diagnosis & Radiology, Govt. Dental College, Mumbai
Correspondence to : Dr S Gupta, E / 32, Ayur Vigyan Nagar, Khelgaon Marg, New Delhi 110049, Telephone No.: 011 26264592

An aspirational biopsy was found to be non productive. An intra oral periapical radiograph showed a multilocular radiolucency associated with a unerrupted right maxillary canine. There was no evidence of any calcification with fine wispy trabeculae arranged in a tennis racket like appearance. There was displacement of roots with loss of lamina dura and root resorption in right maxillary first premolar. The orthopantomograph showed distinct invagination in the floor of the right maxillary sinus. The lesion was found to be corticated except at the alveolar margin (Fig.2). Routine haematological investigations showed slight eosinophilia while serological evaluation of calcium, phosphorus and alkaline phosphatase were found to be normal. An incisional biopsy showed fibrocellular connective tissue with myxomatous changes in some areas suggestive of Odontogenic Fibromyxoma.

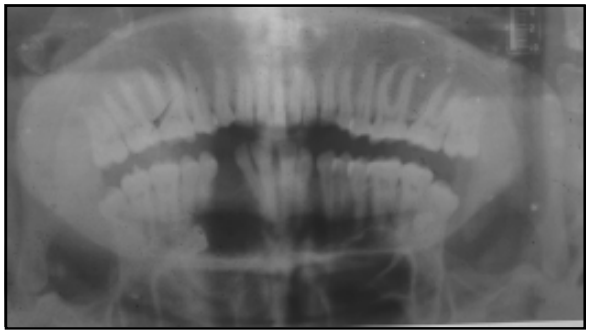

Fig- 2

A CT scan of the maxilla (Fig.3) was carried out at this stage to delineate the true extent of the lesion. This revealed erosion of the cortices in the alveolar margins and a dome shaped extension of the lesion through the floor in the right maxillary sinus.

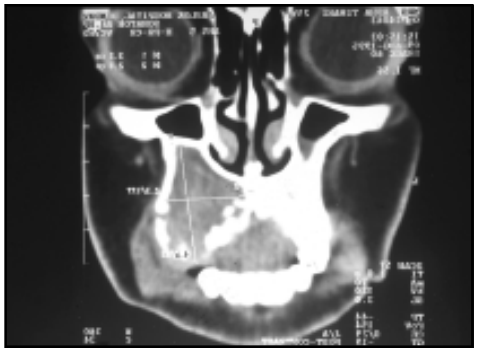

Fig- 3

The tumour was excised and the excisional biopsy confirmed the diagnosis of Odontogenic Fibromyxoma. There were few strands of odontogenic epithelium in the fibromyxomatous connective tissue that was devoid of overlying capsule (Fig.4). There was no evidence of recurrence in the post-surgical follow up at six months and one year.

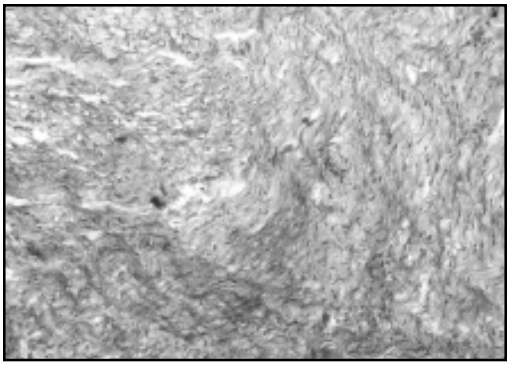

Fig- 4

Discussion

Thoma has described two types of myxoma, Odontogenic myxoma and Osteogenic myxoma, the former he regards as benign while the latter as malignant.(1) Willis regards the so-called Odontogenic myxoma as a fibroma with myxomatous changes while the Osteogenic myxoma as a sarcoma with similar changes. Findings such as limitation of the tumour to teeth bearing areas, its association with unerrupted or displaced teeth and occasional fragments of odontogenic epithelium within the tumour, suggest that it is of odontogenic origin, found exclusively in the bones of the facial skeleton.

Odontogenic Fibromyxoma is a mesodermal tumour representing only about 3% to 6% of the odontogenic tumours.(2) It is a rare, locally aggressive, non-metastizing tumour. It commonly occurs in the second and third decade of life with a slight female predilection and more commonly involving the tooth bearing areas of the mandible (premolar- molar) than the maxilla (4:3). Although some literature refers to this tumour as uncommon in children, it has been suggested that frequency of myxoma in childhood may be higher than that of other aggressive odontogenic tumours(3). Keszler *et al* found 12.5% of myxomas in children with a mean

age of 11.6 years, whereas it has been also reported to occur in a 17 month old child. (3,4) When found in the maxilla it usually involves the zygoma and may invade the maxillary sinus and even the orbits.(5) In the mandible it may involve the neurovascular bundle in the mandibular canal. It is a slow growing painless tumour, which can gradually cause expansion of the cortical plates and cause loosening and displacement of teeth although root resorption may be rare. Only 5% of the tumours may be associated with an unerrupted tooth.(6) If left untreated it can cause perforation of cortical plates, facial asymmetry and regional anaesthesia/ parasthesia.(7) Aspiration cytology is usually not helpful in diagnosis.(8) Radiographically it can appear as a unilocular/pericoronal/multilocular radiolucency or a mixed radiolucent- radiopaque image. Multilocular lesion is more common and larger lesions are more likely to be multilocular.(6) Unilocular lesions are mostly located in the anterior and multilocular in the posterior areas of the jaws.(9) The tennis racket appearance where the bony septae appear radiographically as triangular square or rectangular compartments with very fine trabeculation within them is the most common. The radiographic tumour margins may be either well-defined or poorly defined.(10) Odontogenic myxoma has a variable clinical and radiological appearance and should thus be differentiated from other similar lesions of both jaws in all age groups like ameloblastoma, fibrous dysplasia, intrabony hemangioma, giant cell reparative granuloma and jaw lesion of hyperparathyroidism.(6,10)

The histogenesis of Odontogenic Fibromyxoma is thought to be by one of the following mechanisms-(10)

1. As a direct outgrowth of the dental papilla of a tooth.

2. As an inductive effect of odontogenic epithelium on mesenchymal tissue.

3. As a direct myxomatous change in fibrous tissue.

Microscopically the tumour is made up of loosely arranged spindle -shaped, stellate and round cells with long fibrillar processes that tend to intermesh. The intercellular myxoid substance is composed of two types of acid mucopolysaccharides, Hyaluronic acid (80%) and Chondroitin sulfate (20%). These mucopolysaccharides are believed to be responsible for the neoplastic and aggressive nature of this lesion.(11) Small nests of odontogenic epithelium may be found scattered in the myxoid tissue but are not required for the diagnosis and may not be obvious in all cases. Histologically the tumour must be differentiated from other myxoid lesions like myxoid neurofibroma, myxoid liposarcoma and myxoid chondrosarcoma.(10) A very rare malignant form of the lesion showing marked cellularity and cellular atypism has been designated as myxosarcomas and is found to be more locally aggressive although distant metastases have not been reported.(12) Cytogenetic analysis of malignant myxosarcoma has revealed an unexpectedly aberrant hypertetraploid chromosome complement that was considered as incompatible with the usual karyotypic patterns of benign tumours.(13)

A careful assessment of the clinical, radiographical, tomographical, histological and if necessary immunohistochemical features of the lesion allows a distinction to be made between lesions that are only locally invasive and the rare more dangerous aggressive lesions.(14) Small myxomas are treated conservatively with curettage followed by chemical or electric cautery. Larger tumours may require extensive resection as recurrence rates as high as 25% have been reported and have been attributed to incomplete removal of the original lesion, a problem augmented by local insidious invasion and gelatinous nature of the tissue itself.(7) Wide resection with preservation of vital structures and simultaneous autogenous bone graft reconstruction is preferred.(15) More aggressive surgical treatment should be reserved for lesions for which there is a strong suspicion of malignant transformation.(14) Modern radiographic investigations should be used as an adjunct before surgical manipulation, whenever infiltrative lesions are suspected.(16) Needless to say post surgical follow -up is also a must for such lesions.

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