Case Report

Pediatric Fibromyxoma Maxilla – A rare entity

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ABSTRACT

Fibromyxoma is a rare benign tumour of mesenchymal origin that mostly involves the posterior part of the mandible. It is a locally aggressive and slowly growing painless tumour that mostly occurs in second and third decades of life. We report a case of 2 years old child with huge mass of the right maxilla. After proper diagnosis mass was completely excised via sublabial approach and reported histopathologically as fibromyxoma. Because of its rarity in the maxilla and in this age, it is being reported here.

KEYWORDS: *Odontogenic tumors; Fibromyxoma; Maxilla*

Fibromyxoma is a rare slowly growing benign tumour, which usually occurs in the 2nd and 3rd decades of life. It is rarely seen in children or adults above 50 years of age [1]. Average age of its presentation is 31 years [2]. Fibromyxoma usually occurs in tooth bearing area; thus, is supposed to be arising from the mesenchymal portion of a tooth germ most likely of the dental papilla [3]. It has the tendency of bony destruction and extension into the surrounding structures. Main symptom of fibromyxoma is swelling and problems due to displacement of the surrounding structures [4]. Surgical excision is the mainstay of the treatment. We report this case due to the unusual presentation of this rare tumour in this age involving maxilla.

CASE REPORT:

A 2 year old male child was presented to us in our outpatient department as swelling over right side face for 8 months. Swelling was slowly progressive in size since it was first noticed (**Fig 1**). On examination, there was firm swelling with marked facial disfigurement of right side face. Swelling was non-tender, non-fluctuant and non-pulsatile. Right side proptosis with superior and lateral deviation without any visual impairment was present. Anterior rhinoscopy revealed obliterated right nasal cavity with marked septal deviation towards left side. On oral cavity and oropharynx examination, there was ipsilateral palatal bulge with breach in the mucosa without loosening of teeth. There was no numbness and altered sensations over face or oral mucosa. There was no cervical lymphadenopathy. Rest of the systemic examination was normal.

On investigations, complete blood count and urine examination was normal. Computed Tomography (CT) scan of para-nasal sinuses revealed a large expansile lesion in right maxillary sinus with gross thinning of the bony walls and superior compression over the right orbit. There was hypoplastic frontal sinus with mucosal thickening in the ethmoid, maxillary and sphenoid sinuses more on right sides (**Fig 2, 3**). Previous biopsy from the mass was fibroma.

Child was planned for complete excision of mass under general anaesthesia. After complete pre-anaesthetic work-up, the tumour was approached by means of sublabial route with complete enucleation and curettage (**Fig 4**).

Grossly, the tumour was solid (9cm×5cm×5cm) in consistency (**Fig 5**). Post-operative period was uneventful. Histopathological examination revealed fascicles of benign spindle cells with myxoid area with well-defined capsules suggestive of fibromyxoma (**Fig 6**). There was no recurrence after two year of follow up.

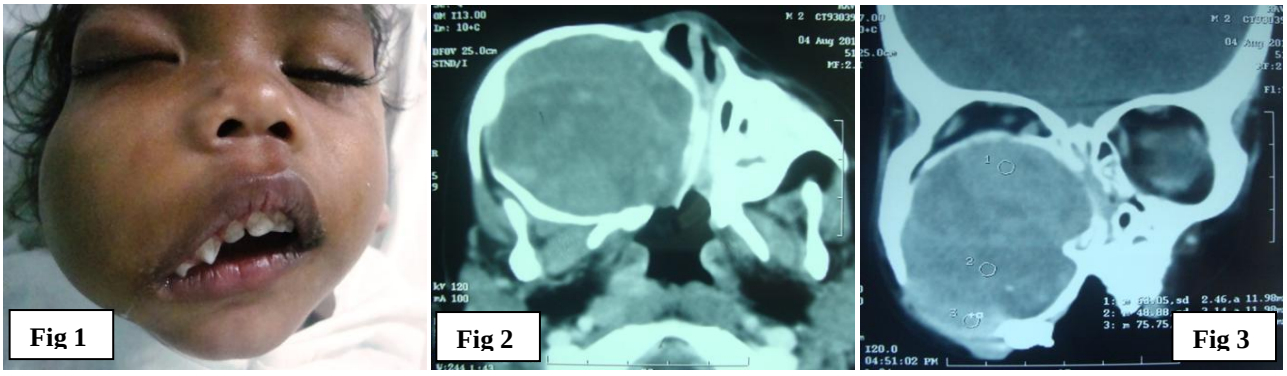

Fig 1: showing swelling over right side face. Fig 2: CT scan axial section showing large expansile lesion in the right maxillery sinus. Fig 3: CT scan coronal section showing large expansile lesion causing gross compression over the right orbit and nasal cavity.

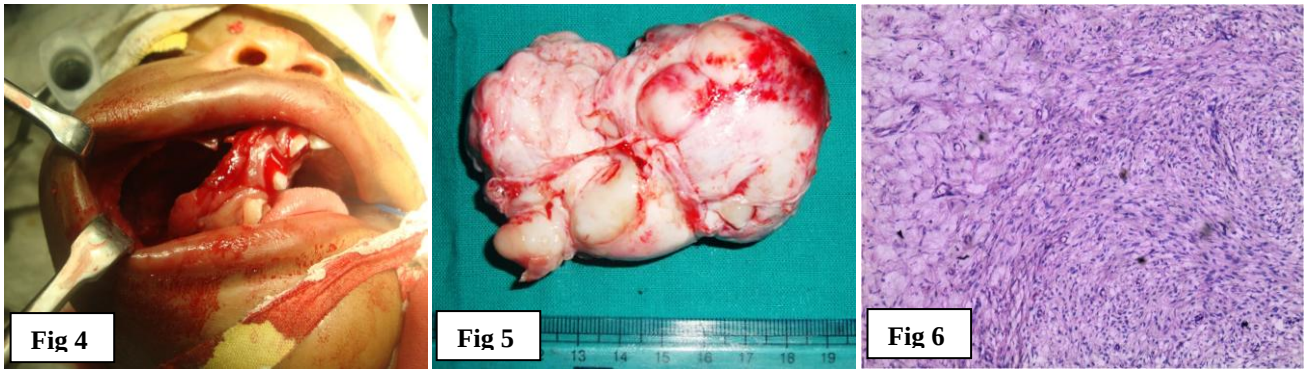

Fig 4: intra-operative photograph. Fig 5: resected specimen. Fig 6: Fascicles of benign spindle cells with myxoid area (H & E 100X)

DISCUSSION

The term myxoma was first used by Virchow in 1863, but the term fibromyxoma was described by Marcove et al in 1964 who reported extragnathic locations of fibromyxoma. In African countries, odontogenic myxoma is the second most common tumour after ameloblastoma (frequency of occurrence 1-19%), while in Asia, Europe and America, frequency lies between 0.5-17%. Overall myxomas represent 2.3-17.7% of all odontogenic tumors with fibromyxomas comprising only a small number of all such tumours [5].

Fibromyxoma is a rare odontogenic tumour with reported incidence rate as low as 0.07 per million population. Fibromyxomas are rare in children under 10 years of age and average age of presentation of this tumour is about 31 years. The tumor is slow growing thus the average delay in presentation is about 3-5 years.

It usually arises from the mesenchymal tissue of the dental papilla; hence, is also known as odontogenic myxoma. The myxoma of the jaw bones is a locally aggressive, non-metastasizing tumor. These are usually located intraorally most often in the posterior region of the mandible, its angle and ramus and rarely extra-orally [6]. The anterior region of the mandible and maxilla are rarely affected. It runs a benign course causing local symptoms leading to loosening of teeth and expansion of alveolus. In early stage the lesion often grows without symptoms; later, there may be marked bony expansion leading to gross facial disfigurement with or without proptosis. Paresthesia,

hyposthesia, anesthesia or pain is relatively uncommon in these patients. Patients with posteriorly located tumours usually present late and with bigger lesions than anteriorly located tumour probably due to more visible disfigurement and malalignment of teeth in lesions of anterior area. Fibromyxoma of the maxilla is more aggressive in nature than that of mandible as is grows through the maxillary sinus.

On gross examination, the tumour appears as a smooth, glistening, gelatinous, lobulated mass with soft to firm consistency [7]. Microscopically, tumour consists of loose stellate cells with long anastomosing cytoplasmic processes [8]. It differs from odontogenic fibroma by absence of collagen fibrous tissue and lesser numbers of odontogenic epithelial rests. A fibromyxoma contains essentially both the features. The radiological examination is done mostly by Computed Tomography, which play an important role in the diagnosis of a fibromyxoma and in the differential diagnosis from other pathological conditions such as ameloblastoma, fibrous dysplasia, aneurysmal bone cyst, giant cell granuloma, intra-osseous hemangioma, simple cysts etc. The radiological features of fibromyxoma are variable and present as uni or multilocular radiolucency with honeycomb and soap bubble appearances [9]. The antral walls are usually expanded but rarely destroyed and tumour margins may differ with the true margins. Fibromyxoma is usually diagnosed on the basis of radiological as well as histopathological examination.

These tumours are managed surgically and the choice of surgical approach and resection is guided by the location and extent of the tumour. Surgery varies from enucleation and curettage to complete resection with or without peripheral osteotomy. The myxomas/fibromyxomas show a recurrence rate up to 25% [10]. The absence of capsule and infiltration in surrounding tissue is the main cause of recurrence. Recurrence can be reduced with more rigorous treatment in the form of partial or complete segmental bone resection. Patient should be followed up at least for 2 years to detect any recurrence of the disease.

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