Recurrent Odontogenic Myxofibroma of the Mandible in a 12 Year Old: An Illustrative Case Report

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Clinical and radiographic features of a large, destructive, unilateral recurrent lesion of mandible in a 12 year old boy histologically proved as myxofibroma are described here. The purpose of this article is to lay emphasis on the importance of early diagnosis of such lesions so that further recurrence can be prevented.
Keywords: *Mandible; Myxofibroma/Myxoma; Odontogenic; Recurrence*
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INTRODUCTION

The term "myxoma/ myxofibroma" was coined by Virchow in 1871, when he described tumors that histologically resembled the mucinous tissue of umbilical cord.[1] In Asia, Europe and America, relative frequencies of myxoma have been reported between 0.5-17.7 percent.[2] It is locally invasive and has a high recurrence rate ranging from 10%-33% with a reported average of 25%.[3, 4]

The World Health Organization (WHO) defines myxoma as a locally invasive neoplasm consisting of rounded and angular cells that lie in an abundant mucoid stroma. The tumor is usually poorly demarcated from the surrounding tissue with which it freely intermingles or from which it is separated by a pseudo capsule.[4] Most tumors termed as myxomas contain little collagen but some consist of isolated thick collagen bands; those are called as myxofibromas.[5] Myxomas of the jaws are believed to arise from the jaw ectomesenchyme and hence all the jaw myxomas are currently considered of odontogenic origin.

A case of odontogenic myxofibroma is presented here in a 12 year old boy describing a brief history and highlighting the main features of a myxofibroma for easy and early diagnosis.

CASE REPORT

A 12 year old boy visited the clinics with the complaint of a large swelling in the oral cavity. The patient gave history of a small lump in the mouth at four years of age for which he was operated upon. No biopsy was performed and therefore diagnosis was not possible at that time. The lesion recurred at the same site and in a span of 8 yrs it had grown to the present size. There was associated pain. On clinical examination a largelump was seen occupying almost the entire oral cavity. The patient had difficulty in speech and mastication. The mass had pushed the tongue to the right side. It was firm on palpation showing mucosal ulceration due to upper anterior teeth (Fig. 1). Ipsilateral submandibular lymph node was tender and mobile and enlarged to a size of 2 cm. Paresthesia was noticed in the region of inferior alveolar nerve.

Orthopantomograph revealed a multilocular, radiolucent (tennis racket) lesion expansile, causing destruction of the cortical bone, producing displacement of the anterior teeth and extending from condyle on side to the anterior part of

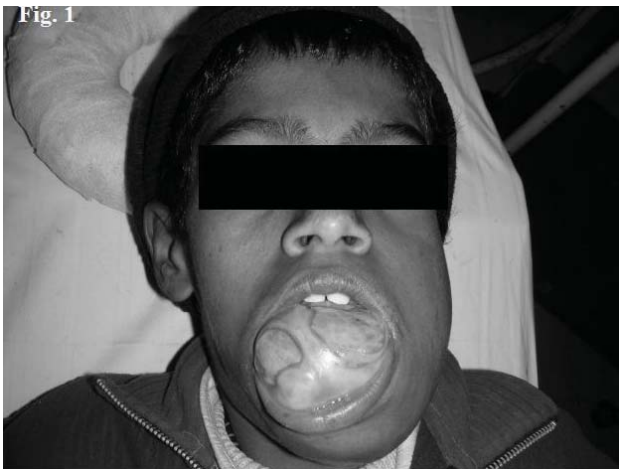

Figure 1. A clinical picture showing the swelling protruding through the oral cavity. The lesion also shows areas of traumatic ulcerations.

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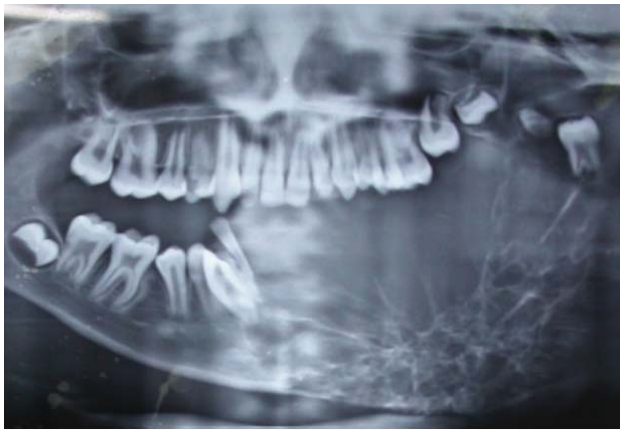

Figure 2. Orthopantomograph (OPG) showing the multilocular, radiolucent (tennis racket) lesion extending from the condyle to the sigmoid notch, expansile, with destruction of the cortical bone and producing displacement of the anterior teeth.

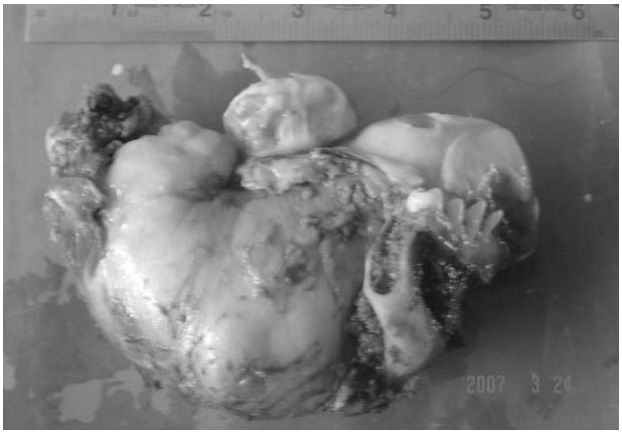

Figure 3. A photograph of the surgically excised huge gross specimen.

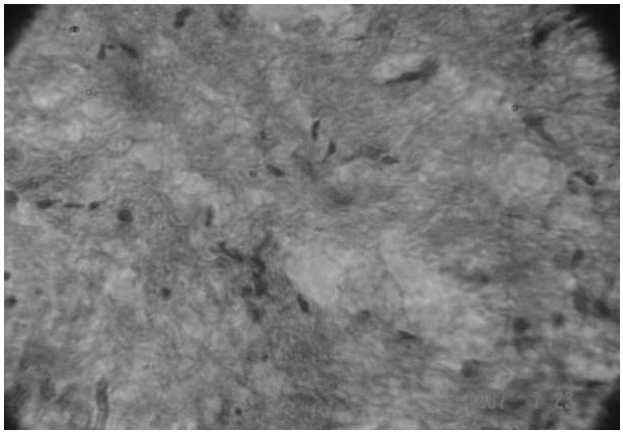

Figure 4. Histopathological picture reveals abundant fibro-collagenous tissue in a myxoid stroma suggestive of myxofibroma. (40X, H/E stain).

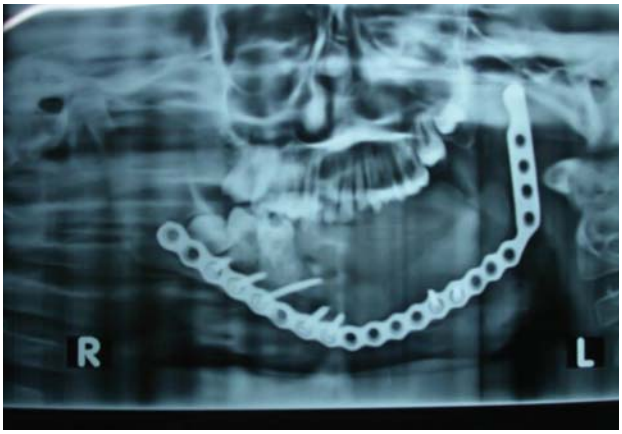

Figure 5. Orthopantomograph of the boy postoperatively showing the fixation of the titanium plate.

mandible with the last molar pushed up to the sigmoid notch (Fig.2).

Histopathological sections revealed abundant fibro collagenous tissue in a myxoid stroma. The cells were relatively uniform in size, stellate in shape and showed no characteristics of malignancy. These cells were evenly spaced within the relatively avascular, fibrillar, mucinous matrix suggestive of myxofibroma. (Fig. 4)

After routine blood examination, the patient was planned for resection with continuity defect, fixation with 2.5 mm Titanium reconstruction plate with condylar head and reconstruction using iliac crest graft. After the surgery, the lymph node subsided on its own. The excised lesion measured about 14x 8x 5 cm and the cut surface was soft, glistening, whitish-grey, translucent and gelatinous with few gritty areas. (Fig.3). Samples of soft tissue from the specimen were processed for light microscopy and these confirmed the biopsy specimen diagnosis of myxofibroma. The patient has been under regular follow-up for the last three years and has not shown any sign of recurrence (Figs. 5–6).

DISCUSSION

Myxoma / Myxofibroma / Fibromyxoma is a non-encapsulated, locally invasive central lesion that does not metastasize and exhibits slow and asymptomatic growth, sometimes resulting in expansion or perforation of the cortices of the involved bone causing visible tumefaction in the mouth.[6] These benign mesenchymal tumors comprise of stellate cells within a mucoid ground substance, and they differ from the myxomas occurring in long bones, which tend to recur and become malignant.[7] The soft tumor mass expands the bone and often causes complete destruction of the cortex.[6,8] Myxofibroma in any form and location is rare. It is considered more uncommon in the jawbones than in the rest of the skeleton.[9] Pure myxofibromas do not metastasize but recur locally if incompletely removed. It is generally agreed that the tumor is locally aggressive and shows tendency to recur and progress if injudiciously attacked. This is more marked in the young before puberty and prior to cessation of the growth as probably was in our case.[10]

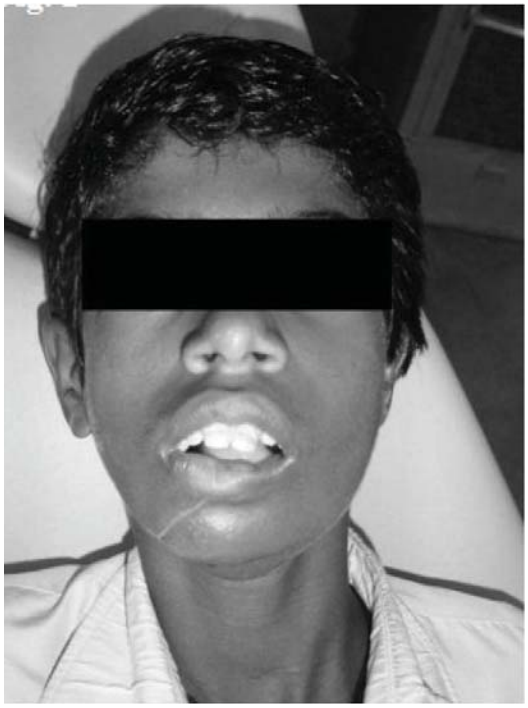

Figure 6. A clinical picture of the boy after the surgery.

In 1948, Stout reported a series of 49 patients with myxomas and established criteria to be satisfied for diagnosis- a true mesenchymal neoplasm consisting exclusively of undifferentiated stellate cells in loose mucoid stroma that did not metastasize.[1,10] The WHO and many authorities consider the jaw myxoma to be an odontogenic tumor on the basis of its site, which is almost exclusively to the tooth-bearing portions of the jaws; the common occurrence in youth or in association with missing teeth; the structural resemblance to dental mesenchyme; and the sporadic presence of islands of odontogenic epithelium.[7]

Thoma and Goldman in their study of 11 cases of myxoma, concluded that the tumor is benign and of odontogenic origin. They consider myxomas of the jaws odontogenic fibromas which have undergone myxomatous degeneration. Stout described a pure myxoma as a true neoplasm composed of stellate cells set in a loose mucoid stroma through which course very delicate reticulin fibers in various directions.[10] They also demonstrated that most of the myxomas in the oral cavity arise from the mesenchymal part of the tooth germ or from retained islands of embryonic tissue.[1,11] The odontogenic variety is thought to arise from the mesenchymal papilla of the developing tooth, either before or after calcification has begun, whereas the osteogenic type can arise from any disturbed mesenchymal focus in the bone. The odontogenic myxoma occurs in the maxilla and the mandible alike. One chief characteristic of the odontogenic lesion is that it occurs in any area where teeth are unerupted or congenitally missing.[6, 9, 11] Willis believed that myxomas and myxosarcomas are merely fibromas and fibrosarcomas in which mucin has developed in the intercellular matrix.[7, 10]

The intra osseous myxofibromas are bone tumors very rarely seen in the facial bones. In the mandible they are primarily solitary, expansive lesions with little predilection for race or sex. They usually occur in patients between 20-50 years of age. The history of the lesion maybe either of complete silence or rapid growth with subjective complaints. Bernier believed that the intra osseous fibroma may well represent the terminal manifestations of the myxoma in which the fiber content overshadows the myxomatous matrix material.[13]

Radiographic features of the odontogenic myxoma are variable, ranging from small unilocular lesions to large multilocular neoplasms, which often displace teeth or, less frequently resorb roots. The multilocular trabecular pattern has been described as honey comb, soap bubble, tennis racket, wispy and spider web in appearance. Most multilocular myxomas are greater than 4.0 cm; unilocular myxomas tend to be smaller. Root displacement rather than resorption is the rule of jaw myxomas.[4,14] *Grossly,* the true myxoma is jelly-like in consistency, whereas the odontogenic myxoma is more rubbery. Myxofibromas are usually well encapsulated and odontogenic varieties may feel gritty on sectioning. It was mono- or multinodular, smooth, shiny, and of a whitish color.[13] In our case too, the lesion was encapsulated and cut surface was soft, glistening, whitish-grey, translucent and gelatinous having few gritty areas.

Microscopically, the true myxoma contains more embryonal fibrous elements while those in the odontogenic myxoma are more mature type. Tumors with both embryonal and mature fibrous elements have thus been called myxofibromas, fibromyxomas, and fibromas with myxomatous degeneration or odontogenic myxoma. The characteristic cell is the small spindle cell with eosinophilic cytoplasm and oval nuclei. Fibromyxoma, a lesion described as having myxomatous degeneration, should be followed for local recurrence.[3, 4, 13] The myxomatous tissue has been described as resembling primitive mesenchyme, the dental papilla and the stellate reticulum.

Treatment of choice is surgical excision. Radiotherapy is avoided in these patients as the predominant histological finding is not one of rapidly dividing cells but one of amorphous intercellular substance, with little evidence of cell division. Numerous types of treatment have been used for these tumors including simple curettage, enucleation, curettage with peripheral ostectomy and en bloc resection with or without immediate reconstruction. Curettage however, seems to be adequate for primary management of the lesions reserving block resection for management of extensive lesions or recurrence.[10, 15, 16]

Thus, based on these findings, once again it is very important that an early diagnosis of odontogenic myxoma/myxofibroma be made so that the problem can be nipped at its bud stage rather than when it aggressively strikes back. Even, if it recurs, the patient and the surgeon should be aware of the fast measures to be taken to make him disease free and both functionally and aesthetically acceptable in the society.

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