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Case Report

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ODONTOGENIC MYXOFIBROMA – A CASE REPORT

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ABSTRACT

Odontogenic myxofibroma is a very rare and locally invasive neoplasm presenting exclusively in the jaw. Most of the cases are asymptomatic, depicted as painless swelling of the jaw. Eventhough they present with varying radiographic features, their diagnosis are always confirmed by the histopathology, exhibiting the classic spindle shaped cells mimicking the embryonal ectomesenchyme in a myxoid background. Here we report a very rare case of myxofibroma, with variable clinical and radiographical presentation.

KEYWORDS: Odontogenic myxofibroma, odontogenic myxoma, odontogenic tumour.

INTRODUCTION

Odontogenic myxofibroma is a very rare, benign odontogenic tumour, derived from the mesenchymal elements of the dental apparatus.[1] WHO defined odontogenic myxoma (OM) as an intraosseous neoplasm characterized by stellate and spindle-shaped cells embedded in an abundant myxoid or mucoid extracellular matrix. The tumour having the capacity to produce some amount of collagen in the same myxomatous background is frequently referred to as myxofibroma.[2] This entity represents only a few percentage of 2.3% to 17.7% incidence of myxomas. Literature searches of this very rare neoplasm revealed only about 24 cases since 1950.[3]

Histologically, myxofibroma is composed of large amount of ground substances which is rich in acid mucopolysaccharides, appearing myxomatous. It also consists of fibroblasts and myofibroblasts, and the ratio of ground substance to cellular content varies among individual cases.[1]

It usually presents as a slow growing swelling, but may lead to cortical expansion and perforation. It is believed to be an aggressive neoplasm, which has local infiltrative potential, accounting for the high recurrence rate. Diagnosis of odontogenic myxofribroma is often found to be difficult, because of its unusual histologic presentation which may look similar to other lesions. But it has to be differentiated from other oral neoplasm because of its aggressive behaviour and recurrence rate.[1,4]

CASE REPORT

A 27 year old male patient presented with the chief complaint of swelling in the right lower jaw for a duration of 7 months. Patient reported that the swelling had gradually increased in time. Swelling was associated with mild pain and discomfort only on mastication. No numbness, paraesthesia, fluid discharge. No history of trauma reported.

On clinical examination, a very mild, diffuse, non-tender, bony hard swelling was noticed on right body of the mandible, extraorally. However, there was no gross facial asymmetry. Surface of the swelling appeared to be normal with no changes in skin texture or colour. On intraoral examination, a solitary swelling was noticed in relation to the marginal and attached gingiva of 83 and 45 in the labial aspect, measuring about 3 x 1 x 1 cm. Colour of the swelling was the same as that of adjacent mucosa (Figure 1). There were retained deciduous 83 and 84 and missing 43 and 44. There was obliteration of the buccal vestibule. On palpation, there was buccal expansion, swelling was firm to hard in consistency and was not tender.

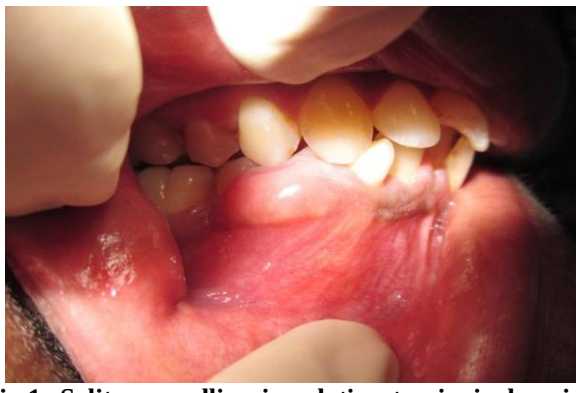

Fig.1: Solitary swelling in relation to gingival region of 83 and 45 with mild obliteration of buccal vestibule.

Orthopantomogram revealed an ill-defined mixed radio-opaque/radiolucent lesion, extending from distal aspect of 32 to distal aspect of 47, migration of impacted 43 to periapical region of 33 and impacted 44 migrated to periapical region of 48. There were osteolytic areas represented by small radiolucent specs. Root resorption was observed in relation to deciduous 83, 84. There was also thinning of the lower border of the mandible (Figure 2).

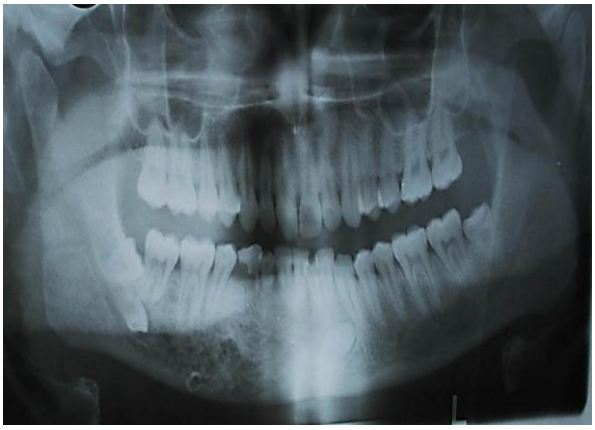

Fig. 2: OPG showing an ill-defined mixed radiolucency extending from 32 to 47, with migrated impacted 43 and 44.

CT axial view does not reveal significant expansion of the right mandible. CT coronal view revealed a mixed ill-defined hyperdense and hypodense regions in the right body of mandible (medullary part) (Figure 3).

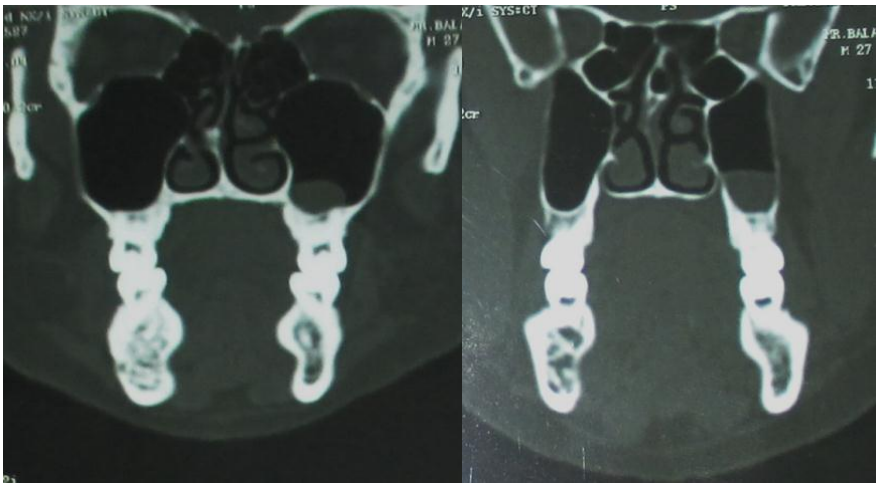

Fig. 3: CT coronal view showing ill-defined mixed hyperdense as well as hypodense region without significant expansion.

Histopathological examination of incision biopsy showed highly fibrous connective tissue without much of vascularity. Most of the areas exhibited pale staining, loose, abundant mucoid stroma which predominantly showed stellate, angular cells (Figure 4 & Figure 5). Some of the areas even showed collagen deposition in the mucoid stroma. Within this background, occasional areas showed islands of odontogenic epithelium. But they were not active-looking in nature. Considering the size of the lesion, and its infiltrative nature, the histopathological diagnosis of Odontogenic myxofibroma was arrived. Patient was advised for the surgical management of the lesion but he was not reported back.

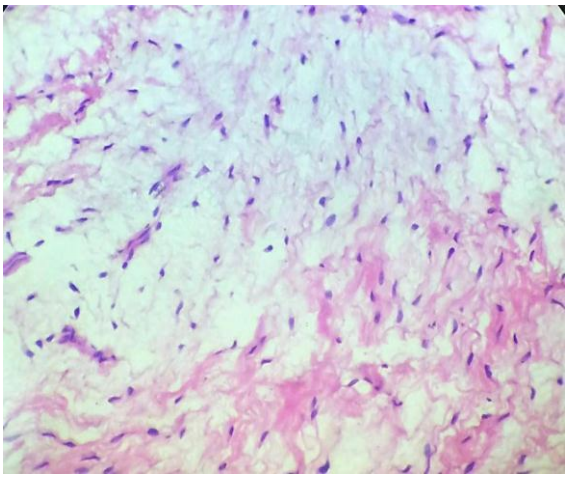

Fig. 4: Photomicrograph showing spindle shaped cells in a myxomatous background, with few collagen fibrils. Also shows collagen bundle formation (H&E 40X)

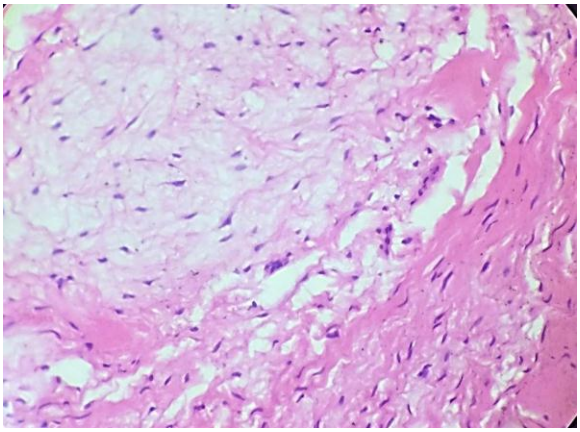

Fig. 5: Photomicrograph showing both spindle shaped cells with both myxoid and fibrous background (H&E 40X)

DISCUSSION

Myxofibroma is a very rare benign odontogenic neoplasm arising from the ectomesenchymal component of the odontogenic apparatus. The term Myxofibroma was first described by Virchow in 1863, who observed this pathological entity had the similar appearance of mucinous substance of the umbilical cord. But Myxoma of jaws was first described by Thoma and Goldman in 1947.[1] Although its frequency is very low comparing them with entire odontogenic tumours, they exhibits distinct histopathological feature with locally invasive potential.

In 1992, WHO defined odontogenic myxoma as 'a benign tumour which is of ectomesenchymal origin with or without the presence of odontogenic epithelium.' It is usually presented as tumour exclusively in the jaw bones, along with unerupted teeth; histologically resembling embryonic dental papilla, dental follicle and periodontal ligament, suggesting their origin; lesion appears as spindle shaped cells, stellate cells or round cells within the loosely arranged myxomatous tissue stroma, with few delicate collagen fibrils. Some cases may show focal presence of odontogenic islands, which appear inactive (i.e. without the presence of tall columnar cells and central space occupied by stellate reticulum like cells). These mucoid tumours have the potential for slow growth, with extensive bony destruction and high recurrence rate if not completely removed. Most of these cases do not present with surrounding capsule, but with gelatinous mucous material at their peripheries.[1]

In this case report, the tumour appeared to be a slow growing lesion, of 7 months duration before the patient seek the examination for the mild facial swelling and intraoral gingival growth. The age range as per literature review indicates that it varies from 22-37 years. It is rare below 10 years and above 50 years. As per location, mandible (66.4%) is considered more common compared to maxilla (33.6%). In the mandible, premolar-molar is the most frequently involved site. The case presented also shows similar age and location as per most of reviewed literatures.

Another frequently associated clinical feature of this entity is missing or unerupted teeth. But in literature, only 5% of cases have been reported with impacted tooth[4]. In our case, there were retained deciduous 83 and 84 and clinically missing 43 and 44. Impacted 43 was found displaced to the left side and placed in relation the periapical region of 32 and impacted 44 pushed to the periapical region of 47 and 48. Impacted teeth were found at the periphery of the lesion, suggesting the tumour has pushed the impacted tooth away from the centre of the lesion. The teeth found in relation to the tumour were often displaced and also shows resorptive features. Similar resorptive features are also noticed in the retained deciduous tooth 83 and 84.

Clinically, odontogenic myxoma are asymptomatic. Pain, paraesthesia or asymmetry occurs only when they present with larger sizes. Typically, these tumours present radiographically as a multilocular lesion; either as honeycomb or soap bubble type. Unilocular lesions are more commonly encountered in children and in anterior part of the jaws. Although most of the tumour present as radiolucent lesion with thin sclerotic borders, some of the cases were reported with mixed radiolucent-radiopaque picture with ill-defined boundaries. Kaffe et al found that mixed radiopaque-radiolucent appearance of odontogenic myxofibroma constitute about 12.5% and radiopaque appearance constitute 7.5%. He further suggested that, myxomas do not have the inherent property to produce calcifications and the radiopacity simply represent the residual bone, entrapped within the neoplasm[4]. In contrast, there are also reports of case of myxofibroma with ossification, in which, it was suggested that those ossified component did not exhibit normal bony architecture. So they suggested that the ossification represent the metaplastic process due to the neoplastic cells.[7]

Cases with mixed radiographic appearance show irregular bony trabeculae with only few septa crossing the centre of the lesion. This produces an overall hazy radiolucent radiologic picture. This hazy appearance is noticed in our case, which can be correlated with the nature of the tumour producing more fibrous component along with the myxoid component. Regarding the borders of the myxoma, Kaffe et al in their review, found that one third of the case exhibited diffuse or poorly defined borders.[4]

Microscopic evaluation of the biopsy tissue is considered to be the most important method to arrive at a definitive diagnosis. Gross specimen exhibiting typical gelatinous, translucent nature of the specimen, un-encapsulated, with ill-defined borders. Histology, typically reveal bland, loosely arranged spindle-shaped, round, stellate cells in a dull eosinophilic cytoplasm, arranged in a mucoid background. These stellate cells may have anastomosing,

angulated, long tapering, cytoplasmic processes. The myxoid matrix is said to be rich in glycosaminoglycans like hyaluronic acid, chondroitin sulphate, which can be demonstrated by Alcian blue at low pH and the fine reticular fibres in the myxoid background can be demonstrated by reticulin stain.[6] Some cases might present with increased fibrous content with collagen deposition, which are referred to as fibromyxoma or myxofibroma. The significance of the presence of the collagen in the myxomatous tumour with respect to their clinical behaviour as well as prognosis is not clear. Based on the these histological features, some of the neoplasm which have the potential for myxomatous degeneration namely fibrosarcoma, chondrosarcoma, liposarcoma can be easily confused, which have different underlying pathogenesis and clinical course.[1]

Other findings include residual odontogenic epithelial islands, which appear inactive and residual bony trabeculae. These unencapsulated tumours are believed to expand by their increasing mucoid ground substances rather than by cellular growth.[5] Expression of vimentin, S-100, muscle specific actin, revealed by most of the studies implicates the origin from mesenchymal component. These findings also correlate with the fibroblastic and myofibroblastic differentiation.[1]

CONCLUSION

Odontogenic myxofibroma, eventhough having a very low incidence, it exhibit differential presentation, especially in its early stage or myxomatous changes within the tumour ground substances. This presentation can be easily confused with other fibrosseous lesion, which are relatively less aggressive and treated in a different approach. So differentiating this rare group of lesion is mandatory with the help of clinical appearance, radiographic findings, and supportive imaging analysis for appropriate management.

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