Odontogenic Fibromyxoma: A Diagnostic Dilemma in a Paediatric Patient

Shimpi Manasi[1], Chaudhary Shweta[2], Kulkarni Venkatesh[3], Chaudhary Mayur[4], Kadam Shraddha[5]

1- MDS III, 2- MDS, Associate Professor , 3- MDS, Professor, 4- MDS, Associate Professor, 5- MDS III

1- Dept. of Paediatric and Preventive Dentistry, Bharati Vidyapeeth Dental College and Hospital, Pune, Maharashtra, India.

***Abstract*:** Odontogenic Fibromyxoma represents a rare slow growing benign neoplasm, found rarely in children below 10 years or adults over 50 years of age. The prevalence of myxoma is between 0.04% and 3.7%. In this case report, a rare case of Peripheral Odontogenic Fibromyxoma in an 8-year-old male has been discussed along with the difficulty in clinically diagnosing the lesion due to its atypical features, which histologically was proved as a Fibromyxoma. The purpose of this article is to discuss the clinical, radiographical and histological features and treatment of this rare benign lesion, laying emphasis on the importance of early diagnosis of such lesions so that further recurrence can be prevented.

Index Terms- *Odontogenic Fibromyxoma, Myxoma, Benign, Neoplasm.*

I- INTRODUCTION

Odontogenic Fibromyxoma is a rare benign, slow growing neoplasm, which usually occurs in the second and third decades of life.[1,2] These are described as a variant of odontogenic myxoma with abundant collagen fibres.[3] Smaller lesions are usually asymptomatic, while larger lesions are often aggressive with possible perforation of the cortical plate.[4,5]

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 tumour is usually poorly demarcated from the surrounding tissue with which it freely intermingles or from which it is separated by a pseudo capsule.[6]

Myxomas of the jaws are rare and are believed to arise from the jaw ectomesenchyme and hence all the jaw myxomas are currently considered of odontogenic origin.
A case of Peripheral Odontogenic Fibromyxoma (POF) is presented here in an 8 year old boy because of its rarity and the diagnostic dilemma encountered while reporting the case. We present a brief history and highlight the main features of a fibromyxoma for easy and early diagnosis.

II- CASE REPORT

An 8 year-old male presented to the Department of Paediatric and Preventive Dentistry, Bharati Vidyapeeth Dental College and Hospital, Pune for evaluation of a soft mass present in the front region of the lower jaw that was a small elevation present at birth and had gradually enlarged since.

The patient reported discomfort when eating during mastication as there was a mobile tooth associated with the mass. He denied any numbness or tingling in the lower lip or chin. He also denied fever, chills, night sweats or weight loss. Medical, dental and family histories were non-contributory. Head and neck examination were negative for any lymphadenopathy.

Intra-oral physical examination demonstrated a sessile, poorly demarcated, expansile mass involving the anterior mandibular region extending from the mandibular primary left lateral incisor till the mandibular permanent right central incisor. The lesion extended buccally across the alveolar ridge to reach the labial vestibule and lingually till the depth of the lingual sulcus. **(Fig. 1)** There was no tenderness upon palpation of the mass. The mass caused displacement of the right mandibular permanent central incisor and left primary mandibular lateral incisor with a grade 1 mobile left mandibular central incisor associated with the mass. The mucosa overlying the neoplasm was intact and no gross ulceration or haemorrhage was observed.

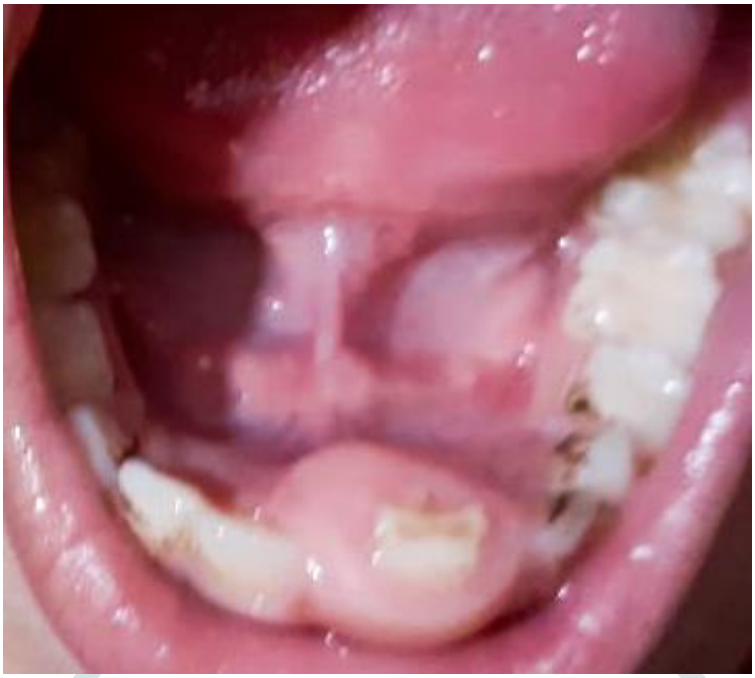

Figure 1- Intra-oral physical examination showing a sessile, poorly demarcated, expansile mass involving the anterior mandibular region.

An intra-oral peri apical radiograph of that region revealed root resorption of the apical third of the left permanent mandibular central incisor. **(Fig.2A)**
A CBCT scan of the region was obtained which revealed an over-retained 71, root resorption with 71,72 and 82 and a missing 31. Loss of the bony crypt and thinning of the labial and lingual cortical plates with respect to 32 and 42 were seen, with incomplete root formation and migration with 41 was seen. A well-defined radiolucency with smooth margin and sclerotic border was seen around the crowns of 32 and 42. The radiographic impression of the CBCT was an eruption cyst with 32 and 42. **(Fig. 2B)**
However, on clinical examination, it was seen that the tooth involved was 31 (not 71) and 32 and 42 appeared to be normal; hence the pathology associated with 32 and 42 was ruled out.

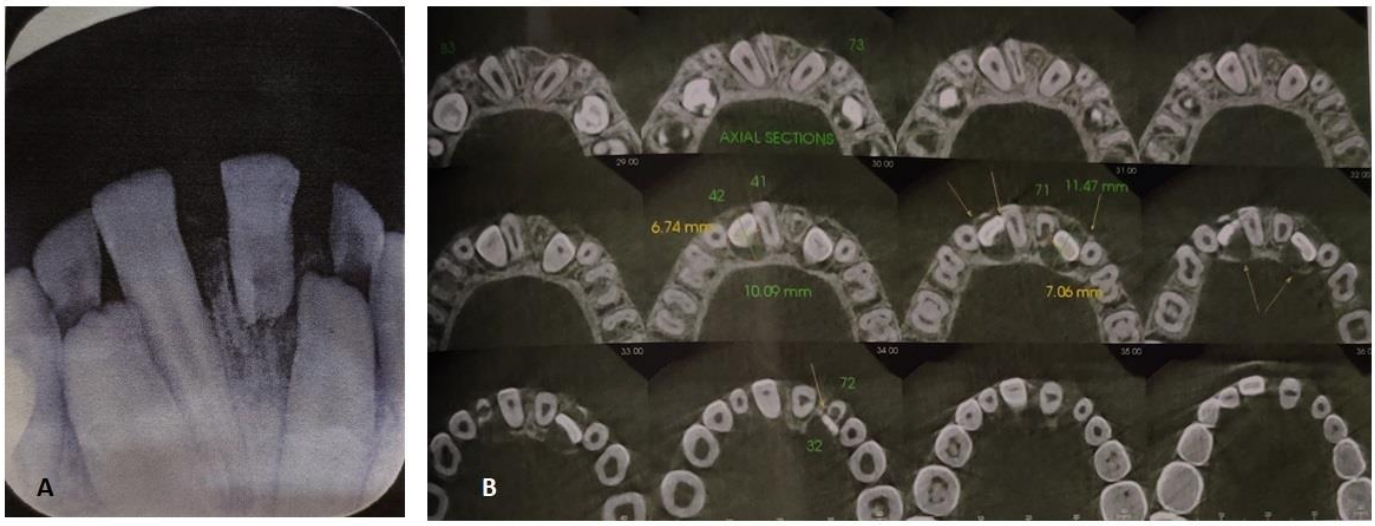

Figure 2- (A) An intra-oral peri apical radiograph revealing root resorption of the apical third of the left permanent mandibular central incisor.

(B) CBCT report showing a well-defined radiolucency with a smooth margin and sclerotic border around the crowns of 32 and 42 giving the impression of an eruption cyst with 32 and 42.

After obtaining an informed consent, extraction of the mandibular left central incisor was done under local anaesthesia (2% Lignocaine hydrochloride) and the patient was kept under observation for regression of the soft tissue lesion. After a 14 day follow-up it was seen that the extraction socket had healed completely but the soft tissue swelling had not regressed. The patient was kept under evaluation for the same for a period of 3 weeks.

At a 1 month follow-up it was seen that the swelling had not reduced in size. **(Fig. 3A,B)** After consulting with oral-surgeons, a provisional diagnosis of an Irritational Fibroma was made and it was decided that the swelling should be surgically removed under local anaesthesia. A mandibular impression was made using Alginate impression material (Zhermack Tropicalgin)

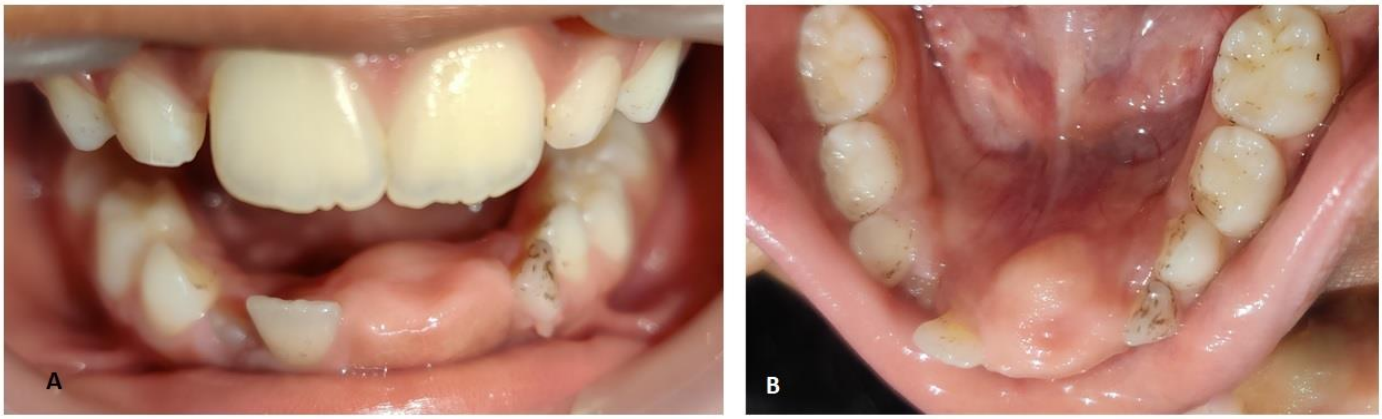

Figure 3- Post- extraction 1 month follow up showing the lesion not regressing in size

After obtaining an informed consent, bilateral inferior alveolar nerve blocks and lingual nerve blocks were given and using electro-surgical cautery, **(Fig. 4A)** the lesion was excised in its entirety till the periosteum, i.e an excisional biopsy was performed and an additional 5 mm of adjacent apparently normal tissue around the lesion was removed keeping in mind that there was no cause for irritation, the lesion was large and diffuse and the commonly seen features of an Irritational Fibroma were not displayed in this case. **(Fig. 4B)** All the tissue tags from the underlying alveolar bone were removed. **(Fig. 4C)** It was seen that there was a slight expansion of the alveolar bone with irregularities seen on the alveolar ridge with sharp margins which were then smoothened using a bone drilling bur mounted in a slow speed hand-piece using a water coolant. A bone file was then used to further smoothen the alveolar bone. The patient was then given a Zinc-Oxide Eugenol Periodontal dressing (GC Coe-pak) following which an acrylic splint was placed extending over the buccal and lingual alveolar ridge till the depth of the sulcus with retentive C-clasps placed with respect to the right and left permanent mandibular molars. The splint was used for covering the ZOE periodontal pack to prevent its displacement. **(Fig. 4D)** Post-operative instructions were given to the patient and the patient was advised a non-spicy semi-solid diet for a period of 2 weeks.

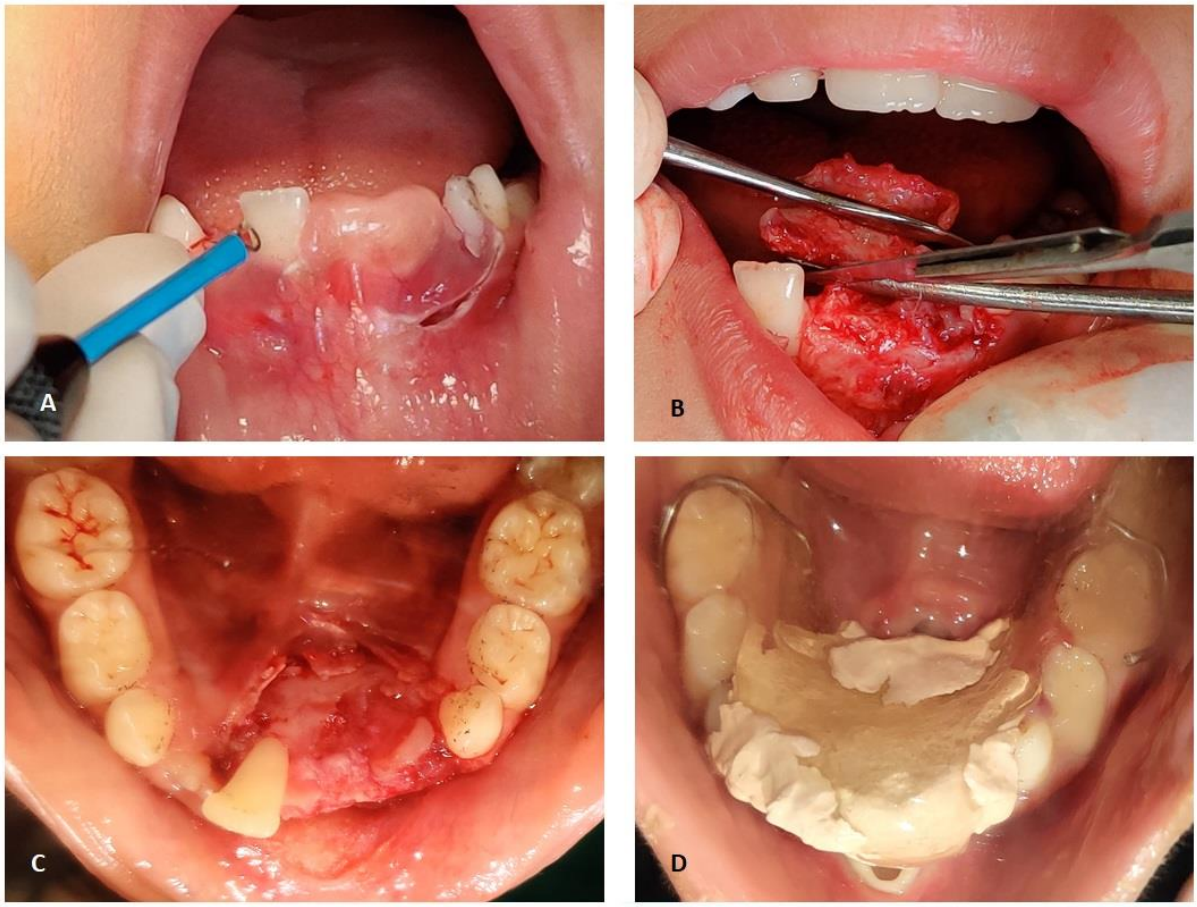

Figure 4 A) Electrosurgical cautery used to perform a wide excisional biopsy
(B) Removal of the lesion in its entirety keeping an adequate safety margin of 5mm
(C) Removal of tissue tags from the alveolar bone followed by smoothening of the irregular bony margin
(D) Placement of a Periodontal dressing stabilised with the help of an acrylic splint

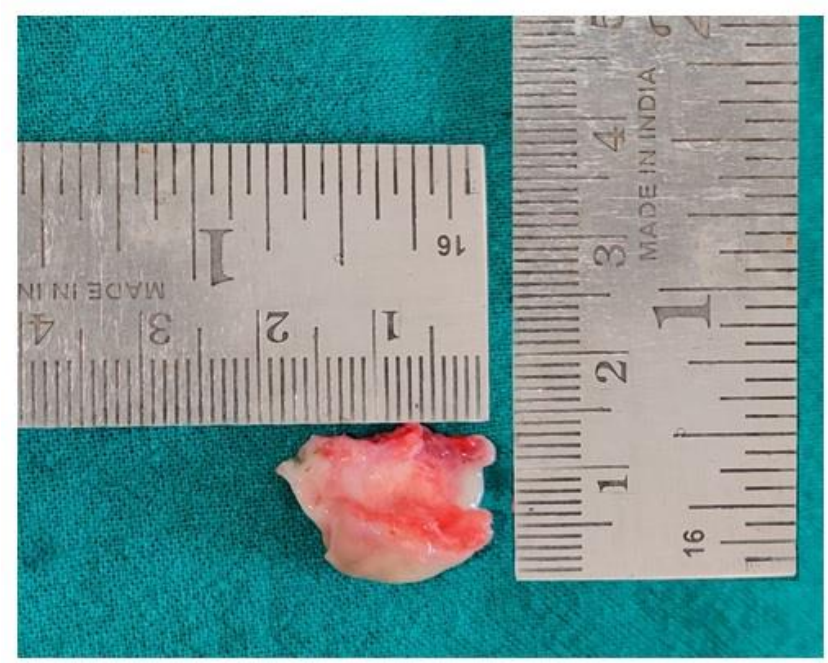

Figure 5- The excised tissue that measured 20mm x 15mm

The excised tissue was placed in a 10% solution of Formalin and sent for Histopathological evaluation. It measured 20mm x 15mm. **(Fig. 5)** The patient was recalled after 2 days for post-operation evaluation. There was no pain or swelling seen with respect to the operating site. However, there was slight tenderness on palpation. The operative site was irrigated using betadine solution and the acrylic splint with the periodontal dressing was placed over the site.

At the 1 week follow up, beginning of re-epithelialisation over the operating site was noted. The periodontal dressing was changed and the acrylic splint was repositioned**. (Fig. 6A)** At the 1- month follow up, tenderness over the lesion had reduced and the patient was advised to continue with a semi-solid diet. The acrylic splint was removed. The underlying tissue appeared to be healing normally. 3 months post-operatively, the surgical site had healed completely and the patient showed no signs of discomfort. **(Fig. 6B)**

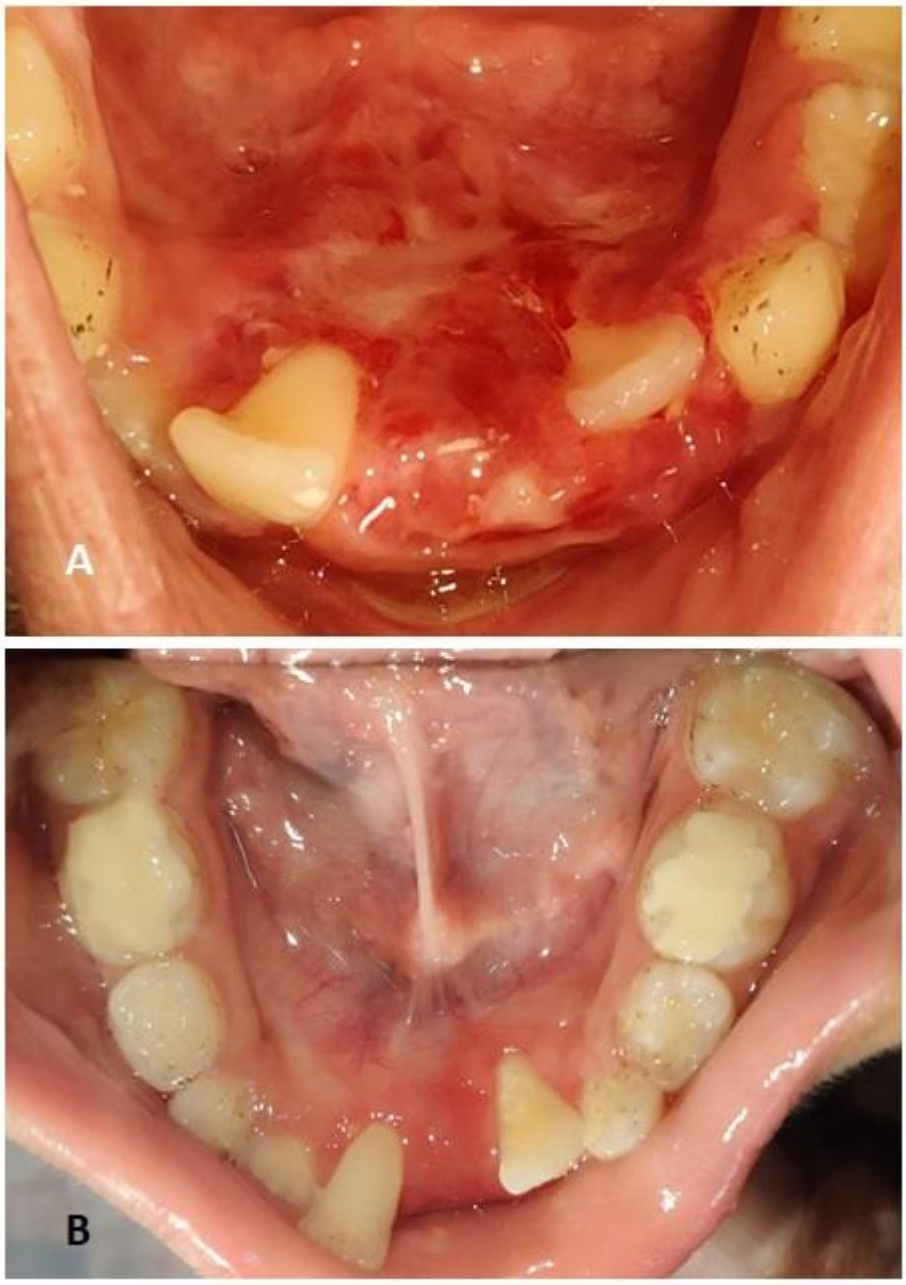

Figure 6 (A) 1 week follow-up image showing beginning of re-epithelialisation
(B) 3 month follow-up image showing showing complete healing

On histological evaluation, the section showed a parakeratinised epithelium lined by stratified squamous cells with irregular rete ridges **(Fig. 7A)**) while the underlying connective tissue consisted of scattered myxoid connective stroma, areas of moderately dense collagen fibers, and islands of odontogenic epithelium. **(Fig. 7B)** The tumour cells had spindle, round or stellate appearances with long anastomosing processes. Nuclear pleomorphism was mild.

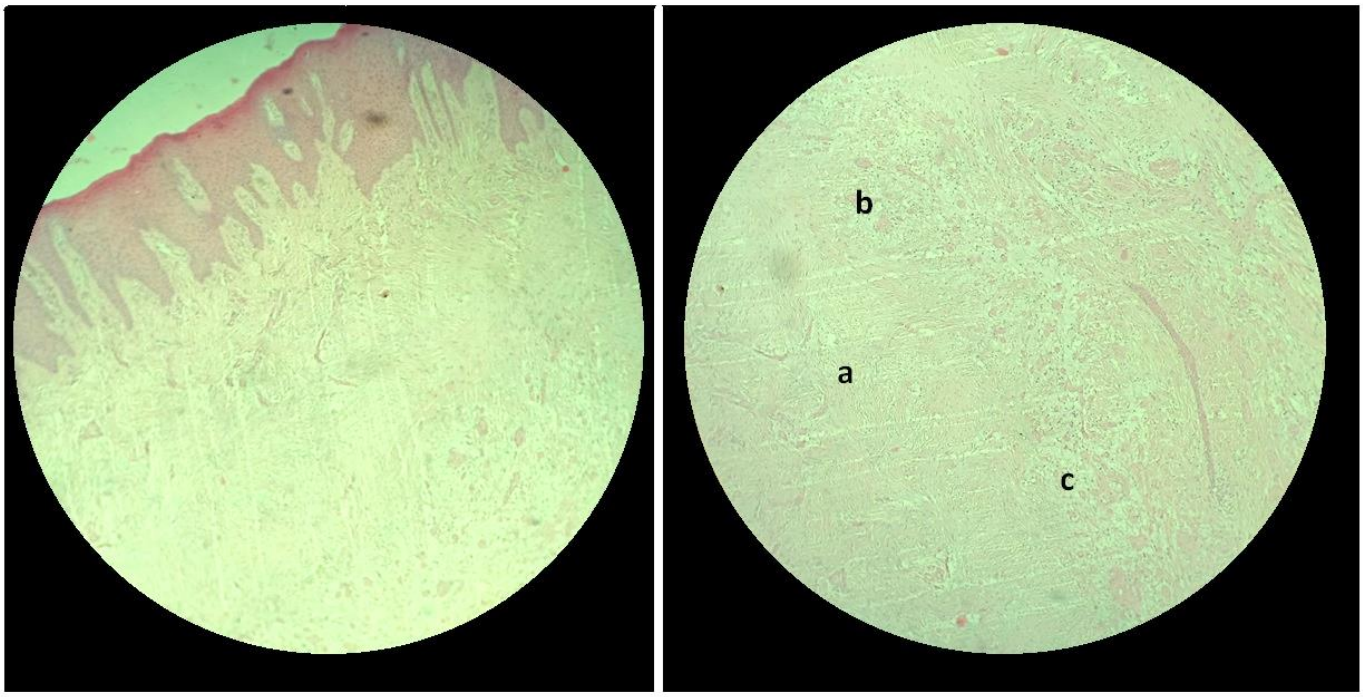

Figure 7 Histopathological investigations

(A) Photomicrography (10x Magnification) showing epithelium and connective tissue stroma
(B) Photomicrography (40x Magnification) showing (a)dense fibrous connective tissue with spindle shaped cells and stellate fibroblasts, (b) odontogenic epithelial islands admixed with (c) myxomatous tissue

III- DISCUSSION

The term "myxoma/ fibromyxoma" was coined by Virchow in 1871, when he described tumours that histologically resembled the mucinous tissue of the umbilical cord.[7]

It is a non-encapsulated, locally invasive central lesion that does not metastasise 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.[8]

In Asia, Europe and America, relative frequencies of myxoma have been reported between 0.5-17.7 percent.[9] It is locally invasive and has a high recurrence rate ranging from 10%-33% with a reported average of 25%.[7,10]A review of literature by Meleti et al has reported only 24 cases of this benign tumour between 1950 and 2015.[5]

3.1 Clinical features-

Fibromyxoma in any form and location is rare. It is considered more uncommon in the jawbones than in the rest of the skeleton.[11] Pure fibromyxomas do not metastasize but recur locally if incompletely removed. It is generally agreed that the tumour 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 [8,12] as probably was in our case.

Grossly, the true myxoma is jelly-like in consistency, whereas the odontogenic myxoma is more rubbery. 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.[8]

In the mandible they are primarily solitary, expansive lesions involving the posterior region of mandible, anterior part of mandible and maxilla are rarely involved.[13,14] with females being affected more commonly than males. [5]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.

Fibromyxomas can be divided into central and peripheral types, with the peripheral type being rarer. In this case, the Peripheral (POF) variety was seen. It is important to determine the type, because it is correlated with the treatment. Fibromyxomas grow slowly without causing any symptoms. Because they enlarge painlessly, they can reach a considerable size prior to being noticed. Lesions can expand the bone, but they perforate the cortex only if they reach a great size. They are locally aggressive lesions, which tend to recur if they are treated too conservatively. Central types can proliferate in the jaw causing bulging of the bone cortex and ultimately breaking through into the surrounding soft tissue. These tumours grow asymptomatically, and loosened teeth may draw attention to the lesions. Otherwise, they are usually diagnosed during routine dental examinations.[15]

3.2 Radiographic features-

Radiographic features of the odontogenic fibromyxoma 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.[6,15,16]

In our case, displacement of 41 and 72 was seen along with resportion of 31 associated with a painless swelling that had enlarged in size since birth.

3.3 Histopathological features-

Histologically, these are characterized by presence of stellate and spindle-shaped cells embedded in an abundant myxoid or mucoid extracellular matrix.[17,18] Trabeculae of woven bone and capillaries, islands of odontogenic epithelium and mast cells are also seen dispersed within the lesion.[5,19]

These benign mesenchymal tumours 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.[8,20] The soft tumour mass expands the bone and may perforate the cortex.[21,22]

3.4 Treatment-

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 tumours 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.[12, 23,24]

The recommended therapy is local conservative enucleation with adequate margins, the extent of which depends on the size and location of the tumour, because the tumour is not encapsulated and its myxomatous tissue infiltrates the surrounding bony tissue without causing its immediate destruction. Consequently, conservative treatment may result in recurrence.[25]

To avoid recurrence, after removing the tumour, all residual tissue tags were removed from the periosteum and the alveolar margin was smoothened off. The patient has been under observation for a period of 2 months and will be further evaluated every 3 months. A conservative approach may be used for smaller lesions to preserve function, reserving rnore radical surgery for recurrences and larger lesions. [15]

IV- CONCLUSION

The present case was unique as it did not highlight the commonly seen clinical and radiological features of POF, due to which it was not considered as a differential diagnosis, proving to be a diagnostic dilemma for the surgeons. Due to the recognition of odontogenic rests along with entrapped bipolar and stellate cells in myxomatous matrix, the final diagnosis of Peripheral Odontogenic Fibromyoma was reached upon. Hence, a triple correlation of clinical, radiological and histopathological findings can guide the surgeon for making appropriate therapeutic decisions.

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