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Case Report

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Fibrous dysplasia affecting maxilla in a 13 year old patient - case report with review

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Abstract

Fibrous dysplasia belongs to a group of fibro-osseous lesions in which the normal bone is replaced by cellular fibrous connective tissue stroma. It is considered as a developmental hamartomatous lesion with cases occurring below the age of puberty. Fibrous dysplasia can occur as monostotic form in which single bone is affected and polyostotic form where multiple bones are involved. Majority of the cases reported are monostotic form with predominant site of involvement being craniofacial skeleton. Polyostotic forms are often associated with MaCune–Albright syndrome, Jaffe–Lichtenstein syndrome and Mazabraud syndrome. The syndromic lesions are manifested with triad of symptoms – fibrous dysplasias, endocrine abnormalities (endocrinopathies like precocious puberty and hypophosphatemia) and skin pigmentations (cafe– au–lait spots). Fibrous dysplasias are expansile lesions and cause complications associated with the site of origin. Maxilla is the most commonest site of involvement in craniofacial skeleton. In this case, a 13 year old male patient presented who was having maxillary fibrous dysplasia.

Keywords: Mazabraud syndrome, fibro-osseous lesions, cherubism, GNAS gene, cafe–au–lait spots

Introduction

Fibro-osseous lesions comprises of a distinct pathological lesions with complications and co–morbid features. Fibrous dysplasia, cherubism, juvenile ossifying fibroma, osteoma and aneurysmal bone cyst are the fibro–osseous lesions commonly encountered in oral cavity. Fibrous dysplasia occurs as relatively rare neoplasm occurring during infancy or childhood. Monostotic fibrous dysplasia accounts for 80–85% of all the cases with jaws being the common site of involvement.[1]

The etiology is mainly genetic with postzygotic somatic mutations of GNAS gene. Mutation of which leads to altered bone forming cells which have fibroblastic phenotype and produce neoplasms at particular sites.[2] Thereby normal bone is replaced by fibrous connective tissue leading to irregular trabecular pattern and woven bone formation. Mostly the diagnosis of the lesion is made by clinical and radiographic features not requiring biopsy. But some lesions pose diagnostic difficulties where biopsy should be performed.

Treatment modalities differ based on the age and clinical behaviour of the neoplasm. Surgical interventions may be difficult as they are more likely to be associated with important anatomical structures. Follow up plays a major role if incomplete resection (remodelling) is done, as the lesions are more likely to recur over time. Bisphosphonate therapy is also indicated in polyostotic fibrous dysplasia. Radiotherapy is contraindicated in these neoplasms as it increases the rate of malignant transformation with frequency of sarcoma occurrence.[3]

In this case report, monostotic fibrous dysplasia of maxilla in a 13-year-old patient is presented.

Case report

A 13-year-old male patient reported to the department of oral and maxillofacial surgery with the chief complaint of swelling in the upper right maxillary region since one year. Patient complained that the lesion was insidious in onset with intermittent growth pattern and attained the present size (Figure 1). The swelling was not associated with pain. No known family history was revealed by the patient. No traumatic history was present. Laboratory investigations were normal. On extraoral examination gross facial asymmetry was noticed on right side of the face, with deviation of nose to left side (Figure 2). Anteriorly the lesion extended below the orbit region at the zygomatic process of the right maxilla without eye involvement. Posteriorly the lesion extended till the malar region of the right side face. Upon inspection the growth was round, circumscribed and well demarcated, measuring 3x5cm. On palpation the lesion was bony hard, non-tender and was not associated with pain. Intraoral examination revealed expansile swelling extending 5cm antero-posteriorly from mesial aspect of 14 to distal aspect of 17 with obliteration of vestibule and 4cm bucco–palatally with bi–cortical expansion, without crossing the mid-palatal line (Figure 3). Retained 54, 55 and clinically missing 13, 15 were observed. Blanching of alveolar mucosa at 55 and 54 was observed. OPG findings revealed radiopaque, ground glass appearance borders blending with adjacent bone (Figure 4). No resorption or displacement of teeth involved was observed. CBCT axial view showed varied degrees of opacifications and coronal view showed thinning of cortical plates, with pushing lateral wall and septum of nose medially (Figure 5 & Figure 6).

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Figure 1 Extraoral photograph of the patient

Figure 2 Deviation of nose is noticed to the left side

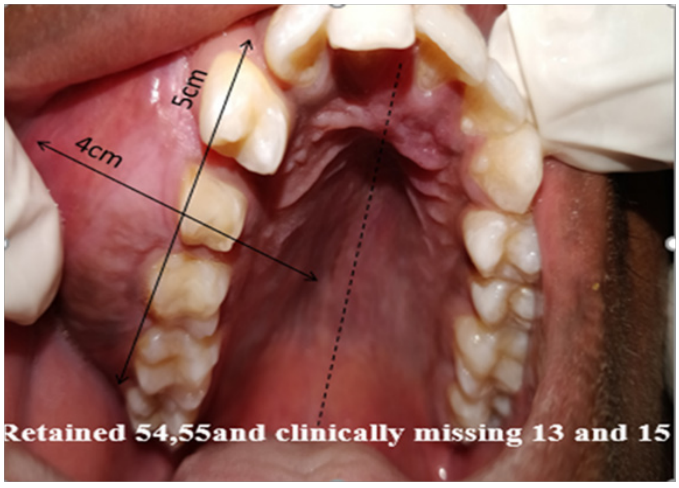

Figure 3 Intraoral Photograph of the patient

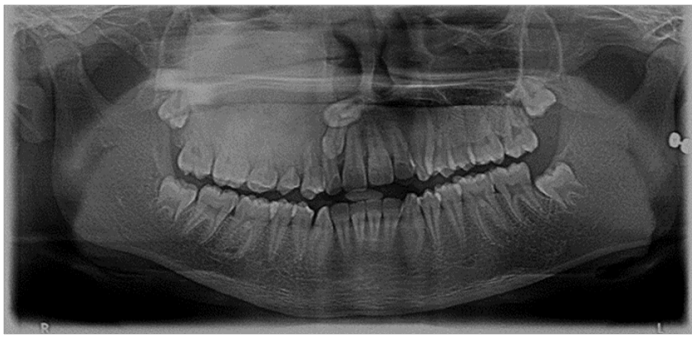

Figure 4 OPG

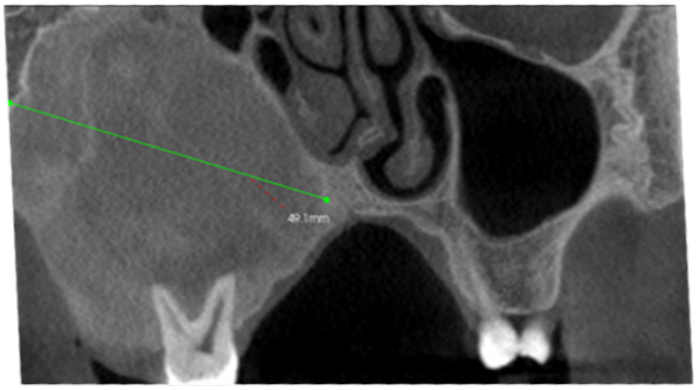

Figure 5 CBCT (axial view)

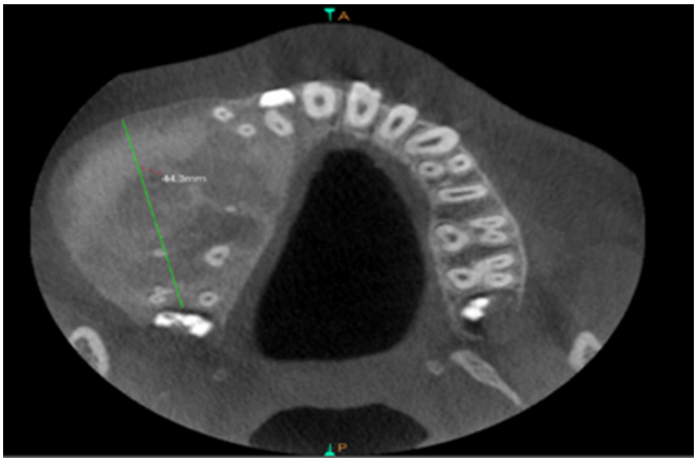

Figure 6 CBCT (coronal view)

Provisionally a diagnosis of fibrous dysplasia was considered. Differential diagnosis of other fibro−osseous lesions like, ossifying fibroma and cemento−osseous dysplasia were considered. Taking into consideration of the age of patient and to retain the teeth involved surgical recontouring and long term follow up was planned. Under general anaesthesia the lesion was shaved, recontoured and reshaped. A vestibular approach was used exposing the anterior aspect till infraorbital region and the lesion was shaved using osteotomes and rotary instruments and while periodically checking the amount of bone removal. After adequate removal of the lesion the area was sutured. The bone bits were sent for histopathological examination (Figure 7).

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Microscopically the H&E stained decalcified sections showed irregularly shaped trabeculae of woven bone in a cellular connective tissue stroma. The bony trabeculae were not rimmed by osteoblasts. Curvilinear shaped trabeculae were evident in focal areas. Lesional bone was fused with the normal bone. Artifactualperitrabecularclefting was also noticed. The fibrous stroma showed coarse, irregularly arranged collagen fibers with fibroblasts (Figure 8). Based on clinical, radiographic and histological features a final diagnosis of monostotic fibrous dysplasia of maxilla was given and patient was advised long term follow up.

Figure 7 Grossing picture of the lesion after biopsy

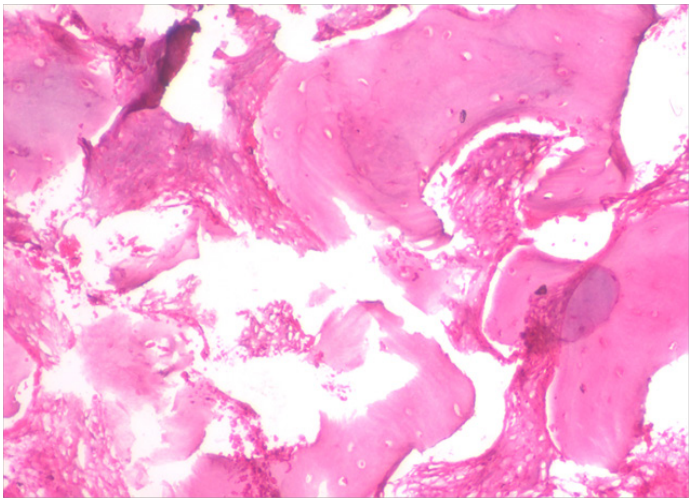

Figure 8 H&E picture of the lesion

Discussion

Fibrous dysplasia is a benign congenital fibro−osseous lesion with normal bone replaced by fibrous tissues. It was first described by Albright et al in 1937 in a patient with syndromic symptoms of skeletal neoplasms, skin pigmentation and endocrine abnormalities.[4] Pathogenesis of fibrous dysplasia mainly involves mutations of GNAS1 gene (Guanine nucleotide-binding protein) located at chromosome 20q13.2.[5] Early postzygotic mutations cause polyostotic fibrous dysplasia with multiple sites of involvement and late postzygotic mutations cause monostotic fibrous dysplasia with single site of occurrence. As these mutations are somatic familial history is not expected, as seen in present case. These mutations cause hyperfunction of bone progenitor cells which acquire fibroblastic phenotype. The medullary and cortical bones are replaced by sheets of fibro−osseous tissues with intermixed trabecular bone. Fibrous dysplasia occurs below the age of ten years with no gender predilection.

Clinically the lesion exhibits severe asymmetry and associated symptoms like visual impairment, diplopia, proptosis, hearing loss, nasal obstruction, epistaxis, epiphora, pain and paresthesia.[6] Laboratory investigations show increased levels of alkaline phosphatase, serum calcium and serum phosphorous levels. But in monostotic forms the laboratory investigations are normal like in the present case. Radiographic features are generally diagnostic with characteristic ground glass appearance or orange peel or cotton wool appearance of bony trabeculae. Displacement and root resorption of associated teeth is absent in fibrous dysplasia as seen in present case. Sometimes the density of fibrous dysplasia mimics multilocular appearance often misleading the diagnosis as ameloblastoma. Differentially diagnosis of ameloblastoma, ameloblastic fibroma, ameloblastic fibro-odontoma, central giant cell granuloma, odontogenic cyst, ossifying fibroma, osseous dysplasia, chronic sclerosing osteomyelitis and osteosarcoma are considered for fibrous dysplasia. Treatment may involve radical surgery for devastating forms or normal shaving and debridement as a part of conservative treatment. Sometimes the lesion is said to be regressed as the patient reaches adulthood, but this concept is not proven. Few case reports also mentioned about deferring treatment till the patient reaching above ten years of age or as such with a close follow up.

Conclusion

In our case report, we presented clinical, radiographic, histopathological features and treatment plan for a case of monostotic fibrous dysplasia of maxilla. Proper clinical and radiographic features of the patient are mandatory for confirmative diagnosis. Based on the clinical behavior and age of the patient appropriate treatment has to be planned with early intervention to avoid complications. Recurrences are common for fibrous dysplasia, hence long term follow up should be done. Our case with its clinical and radiographic features represents an addition to the literature of monostotic fibrous dysplasia.

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