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Monostotic Fibrous dysplasia of mandible: A Case Report

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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. It is characterised by a blend of fibrous and osseous elements in the region. It is a lesion of unknown etiology, uncertain pathogenesis, and diverse histopathology. With an incidence of 1:4000-1:10,000 it seems to be a rare disease. It represents approximately 2.5% of all bone lesions and about 7% of all benign bone tumors. 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. This report describes a case of fibrous dysplasia of a thirteen year female patient who had unusual presentation involving right mandible. The clinical findings, radiological findings and treatment have been discussed.

Introduction

Fibrous dysplasia belongs to a group of fibro-osseous lesions in which the normal bone is replaced by cellular fibrous connective tissue stroma. 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. Fibrous dysplasia can occur as monostotic form in which single bone is affected and polyostotic form where multiple bones are involved.Monostotic fibrous dysplasia accounts for 80−85% of all the cases with jaws being the common site of involvement.[1] It was first described by Albright et al in 1937 in a patient with syndromic symptoms of skeletal neoplasms, skin pigmentation and endocrine abnormalities.[2] FD is caused by somatic activating mutations of the gene GNAS in a subunit of the stimulatory G protein, located at 20q13.2-13.3.[3] Treatment of bony lesions of fibrous dysplasia includes surgical and nonsurgical therapies. Surgical treatment in young-aged minor cases and biopsy with minor bony osteoplasty at affected site are adequate. In more severe cases complete excision with graft reconstruction may be possible.[4] 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.[5] Monostotic FD (MFD), although less serious than Polyostotic FD (PFD), is of greater concern to the dentist because of the relatively high frequency of occurrence in the jaws.[6] In this case report, monostotic fibrous dysplasia of mandible in a 13-year-old female patient is presented.

Case Report

A 13-year-old female patient reported to the department of oral medicine and radiology with the chief complaint of swelling in the lower right mandibular region since one year. Patient complained that the lesion was insidious in onset with intermittent growth pattern and attained the present size. The swelling was not associated with pain. There was no history of trauma, trismus, diminished vision or loosening of teeth. No known family history

was revealed by the patient. Clinically the patient presented with facial asymmetry. Extraoral examination revealed facial asymmetry with well-defined bony hard swelling in the right mandibular region. (FIG: 1) On palpation the lesion was bony hard, non-tender and was not associated with pain. Intraoral examination revealed expansile swelling extending antero-posteriorly from mesial aspect of 44 to distal aspect of 47 with obliteration of vestibule due to buccal cortical expansion. (FIG:2) There was no swelling present elsewhere in the body, and café-au-lait spots were absent.

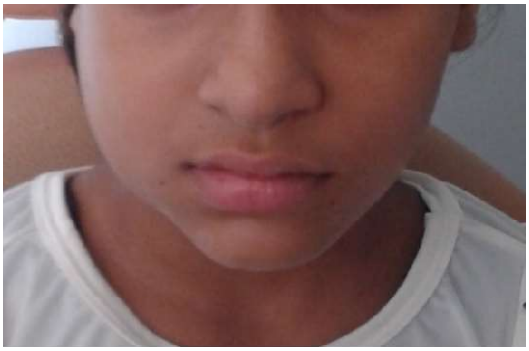

Figure 1: Extra oral photograph showing facial asymmetry on the right side

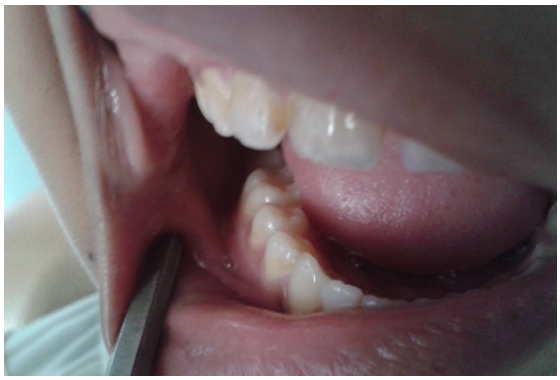

Figure 2: Intraoral photograph showing expansion of buccal cortical plate on the right side

Routine investigations such as hemogram, serum calcium, .serum phosphorus and serum alkaline phosphatase (ALP) were performed. All parameters were within normal limits. An Orthopantomogram (OPG) revealed a poorly defined mixed density pattern giving ground-glass appearance in the right mandibular angle region. (FIG:3) No resorption or displacement of teeth involved was observed. Other features like maxilla, mandibular symphysis, parasymphysis, body and ramus of the mandible appeared to be normal. Bilateral glenoid fossae and articular eminences grossly showed no remarkable changes. Bilateral condylar heads also appeared normal.

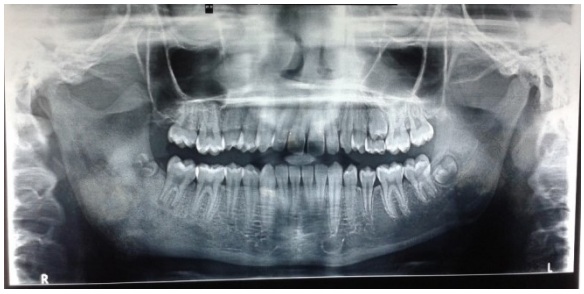

Figure 3: Panoramic radiograph showing mixed radiopaque and radiolucent lesion on the right side of the mandible

Provisionally a diagnosis of fibrous dysplasia was considered. Differential diagnosis of other fibro−osseous lesions like, ossifying fibroma and cemento−osseous dysplasia were considered.An incisional biopsy was obtained from the lesion in the mandible and histopathological analysis was done. Macroscopy showed one piece of bony hard-tissue, irregular in shape, hard in consistency, creamish white in color measuring 0.8x0.6cm approximately (FIG:4)Microscopically the H&E stained decalcified section showed irregularly shaped trabeculae of woven bone in a cellular connective tissue stroma. Bony trabeculae showed no osteoblastic rimming. Poorly formed metaplastic bone was separated by cellular fibrous connective tissue stroma.Curvilinear shaped trabeculae were evident in focal areas. Lesional bone was fused with the normal bone. Artifactualperitrabeculareclefting was also noticed. The fibrous stroma showed coarse, irregularly arranged collagen fibers with fibroblasts (FIG:5)

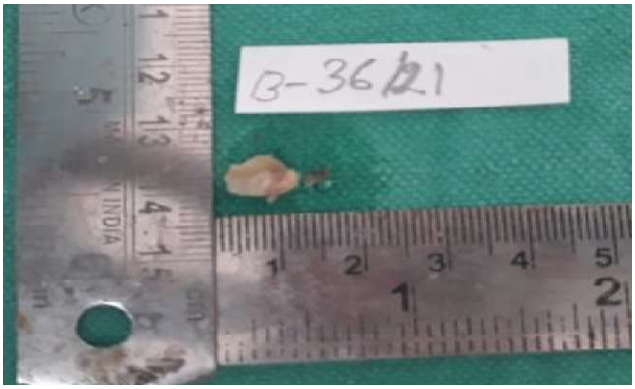

Figure 4: Grossing picture of the lesion after incisional biopsy

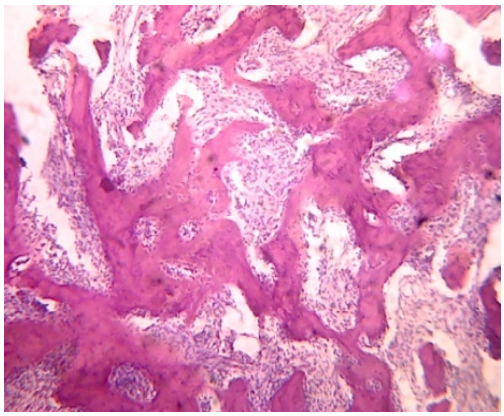

Figure 5: Decalcified section (H&E X10) showing irregularly shaped trabeculae of immature bone within proliferating fibroblastic tissue.

Based on the clinical history, radiographic assessment and histological features of the lesion, a diagnosis of monostotic FD was deduced. Surgical recontouring of the mandible along with contour correction of the alveolar bone was performed. Patient was discharged and regularly followed up at 1 week, 1 month, every 3 months up to 3 years, and then once every year till the 5th year.

Discussion

Lichtenstein [7] first described fibrous dysplasia (FD) as a progressive congenital disorder which is characterized by intermixed, thickened, and woven growth of immature and fibrous tissue with normal cancellous bone.[8] It may arise as a single lesion referred to as monostotic or can occur with multiple lesions that affect many bones also known as polyostotic. A small set of polyostotic FD can also occur as a component of a multisystem developmental disorder known as McCune-Albright syndrome that is also associated with endocrine hyper function and café au lait cutaneous macules.[9]MFD is most common, with a distinctive male predilection showing 70% of cases mostly in puberty and up to 50% female predilection in PFD, which involves craniofacial bones. It is usually observed in children and young adult patients, with 75% of them presenting before 30 years of age. FD is caused by somatic activating mutations of the gene GNAS in a subunit of the stimulatory G protein, located at 20q13.2-13.3.3The GNAS1 gene encodes a G-protein that stimulates the production of cAMP. The mutation results in the continuous activation of the G-protein leading to overproduction of cAMP in affected tissues. Hyper functioning of affected endocrine organs frequently gives rise to precocious puberty, hyperthyroidism, growth hormone excess, and overproduction of cortical. There is increased proliferation of melanocytes resulting in large café-aulait spots. cAMP is thought to have effect on the differentiation of osteoblasts leading to FD.[10] Laboratory investigations show increased levels of alkaline phosphatise, serum calcium and serum phosphorous levels. But in monotonic 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. The differential diagnosis of FD includes other fibro-osseous lesions such as cherubism, juvenile ossifying fibroma, osteoma and aneurysmal bone cyst.Florid FD, Aneurysmal bone cyst, Giant cell granuloma, are often confused with FD. The gold standard for confirming the diagnosis of FD is a histologically proven fibro-osseouslesion radiographic finding with poorly defined margins. “FD is a bony developmental anomaly showing proliferation within the medullary bone as hamartomatous fibrous tissue, producing newly immature, weak calcified bone, with secondary bony metaplasia and without any maturation of osteoblast which on radiographs appear as radiolucent, described classically as ground-glass appearance.”[11] No specific treatment exists for FD. Radical resection is the only technique to obtain complete resolution of FD. Regular recall and follow up is indicated in cases of stable lesions and the lesion usually ceases to grow once the patient reaches puberty. Reconstructive techniques allow obtaining adequate aesthetical and functional results. Aggressive lesions are treated by radical resection, except in pediatric patients with residual large defects in which it is acceptable to try to resolve symptoms through bone shaving, reserving more aggressive treatments in cases of relapse or after skeletal maturity.[12] Medical treatment with bisphosphonates may have benefits including improvement of function, pain relief, and lower risk of fractures in properly selected FD patients.

Conclusion

In our case report, we presented clinical, radiographic, histopathological features and treatment plan for a case of monostotic fibrous dysplasia of mandible. Isolated FD cases in maxillofacial regions are few and are not very easy to differentiate from other disorders of benign and malignant bone origin.

Imaging studies and histological and laboratory tests are very essential to be carried out for gaining definite diagnosis, treatment planning, and for management of FD. Based on the clinical behaviour and age of the patient, early intervention and appropriate treatment should be planned to avoid complications. Recurrences are common for fibrous dysplasia; hence long term follow up is mandatory.

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Conflicts of interest

There are no conflicts of interests

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