CASE REPORT

Craniofacial fibrous dysplasia

Aakarsh Jhamb, Sujata Mohanty[1], Parul A Jhamb
Department of Oral and Maxillofacial Surgery, Santosh Dental College and Hospital, [1]Maulana Azad Institute of Dental Sciences, New Delhi, India

Address for correspondence:
Dr. Aakarsh Jhamb,
SFS 111, Rajouri Aptts, Rajouri Garden,
New Delhi-110 064, India.
E-mail: draakarsh@gmail.com

ABSTRACT
Fibrous dysplasia can present clinically in varied forms which may appear as collision of different pathologic processes. We report a rare case of craniofacial fibrous dysplasia with coexisting epithelial lined cyst and superimposed osteomyelitis with sequestrum formation. Its clinical features and management with possible hypotheses are described along with the post operative course. Pertinent literature has been reviewed with emphasis on pathogenesis of this unique occurrence.
Key words: Craniofacial fibrous dysplasia, epithelial lined cyst, osteomyelitis

INTRODUCTION

Fibrous dysplasia is a congenital developmental anomaly of bone which may occur in localised, regional or systemic forms. In this condition the normal bone is replaced by immature fibro cellular tissue, which may be considered to be a fertile soil for a plethora of lesions like aneurysmal bone cyst,[1-5] solitary bone cyst,[6,7] and non specific cystic degeneration.[8-10] However, the occurrence of an epithelial lined cyst in the maxilla affected by fibrous dysplasia has never been reported. We present a rare case in which an epithelialised inflammatory cyst lining was present along with sequestrum in the maxilla affected by fibrous dysplasia.

CASE REPORT

A 24 yr old man reported in March 2007, with complaints of swelling and pus discharge from right side of cheek since 3 weeks. The patient was apparently well 8 years back when he first noticed slight asymmetry of right cheek region. The swelling slowly increased in size for next three years and was latent since then (about 5 years). Recently, he had toothache in relation to right maxillary molar teeth and noticed pus discharge from the same quadrant. This prompted him to seek treatment and was then referred to our institution by a general physician. He appeared to be a well built young man with no history of any systemic diseases. There was an obvious swelling over the right side of maxilla extending over the body of zygoma. The infraorbital rim was expanded but ocular level in the vertical and horizontal plane was unaltered.

On intraoral examination, a full complement of teeth was present in the right maxillary quadrant. Teeth #17 was grossly carious with a small sinus tract in the alveolus of tooth #15 and 16. On palpation, the swelling was hard, non tender and appeared to blend with the right zygoma and uninvolved surface of right anterior maxilla [Figure 1]. There was no paresthesia over the distribution of infraorbital nerve. Conventional Radiographs showed haziness and opacification over the region of right maxilla and zygomatic bones, containing an ill-defined radiolucency with another small central radiopacity anteriorly. Overall the radiopacity was homogenous and generally appeared to blend with the adjacent bone except for that portion of radiolucency circumscribing another small central diffuse radiopacity. CT scan showed enlargement of right zygomatic and maxillary bones with a diffuse ground glass appearance obliterating the maxillary sinus [Figure 2]. There was an irregular small cavity in the alveolar process of right maxilla around the roots of teeth #15 and 16. A stellate shaped bony sequestrum was seen inside this cavity with a sinus opening on the buccal surface of tooth #16 [Figure 3].

Laboratory investigations like serum alkaline phosphatase, serum calcium, phosphorus and complete blood count were normal. A clinical diagnosis of osteomyelitis superimposed on fibrous dysplasia of right maxilla and zygoma was formed. It was decided to perform facial recontouring and curettage with extraction of involved teeth as a single stage procedure. After a brief course of antibiotics the patient was scheduled for surgery under local anesthesia.

Access this article online
Quick Response Code:
Website: www.jomfp.in
DOI: 10.4103/0973-029X.102523

Operative procedure

Under right maxillary nerve block supplemented with local infiltration for haemostasis, a mucoperiosteal flap was raised and the lesion was exposed till the infraorbital rim superiorly and body of zygoma posterosuperiorly. In the alveolar region the buccal cortex was destroyed and replaced by thick cystic lining. Teeth #14-17 were extracted and cystic lining was curetted out. The cystic lining contained little bit of pus and three small pieces of necrotic bone. Then the residual bone was curetted particularly over the anterior surface of maxilla, body of zygoma and infraorbital rim [Figure 4]. Since there was no paresthesia or compression symptom in infraorbital nerve, the rim of bone surrounding it was left untouched.

On histopathology, the bony pieces retrieved after recontouring revealed uniformly distributed curvilinear trabeculae in a fibrocellular stroma with numerous fibroblasts and a few blood vessels, overall picture appeared to be consistent with mature stage of fibrous dysplasia [Figure 5].

The lining exhibited a nonkeratinised stratified squamous epithelium with proliferating and arcading rete ridges and an underlying connective tissue capsule showing intense infiltration by chronic inflammatory cells chiefly lymphocytes and plasma cells [Figure 6]. The three pieces of bone contained within the lining were necrotic i.e. sequestrum [Figure 7]. Thus a diagnosis of osteomyelitis with coexisting epithelialised cyst in fibrous dysplasia was confirmed.

One year postoperatively good symmetry was maintained in the frontal view but slight expansion was still visible on the worm's eye view. No recurrence of cystic cavity was observed in one year post operative CT scan and the bony defect healed completely. Three years postoperatively the patient came back seeking correction of facial asymmetry caused by bony growth in the same region [Figure 8]. Facial recontouring was performed again and the bone pieces removed showed microscopic picture consistent with fibrous dysplasia. No cystic lining or avascular bone was observed.

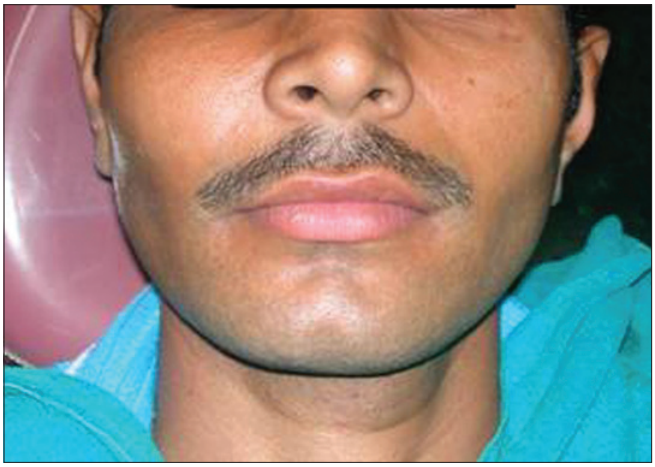

Figure 1: Preoperative photograph showing swelling over right zygomatic region

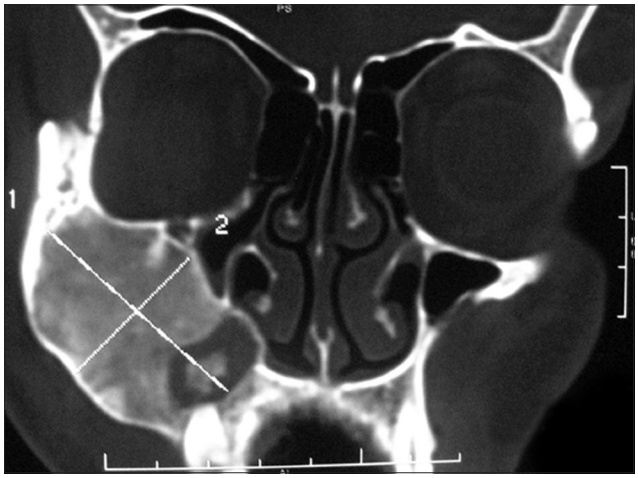

Figure 2: CT scan coronal section showing the extent of lesion with typical ground glass appearance and cystic lesion with sequestrum. The cystic lining appears to be in continuity with root apex

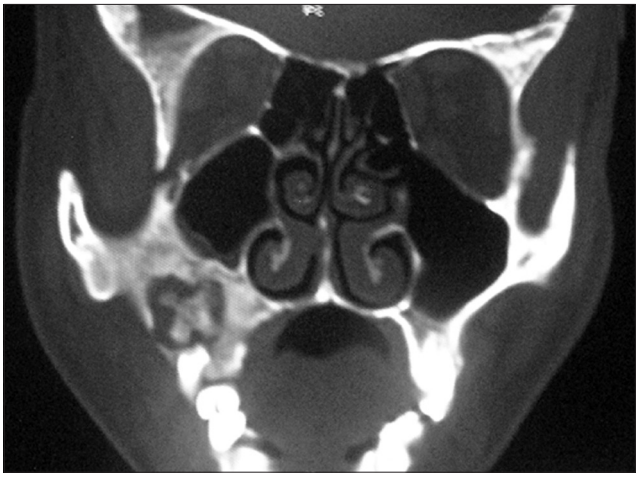

Figure 3: CT scan showing stellate shaped sequestrum surrounded by cystic lining and perforation in the lining wall towards the buccal cortex

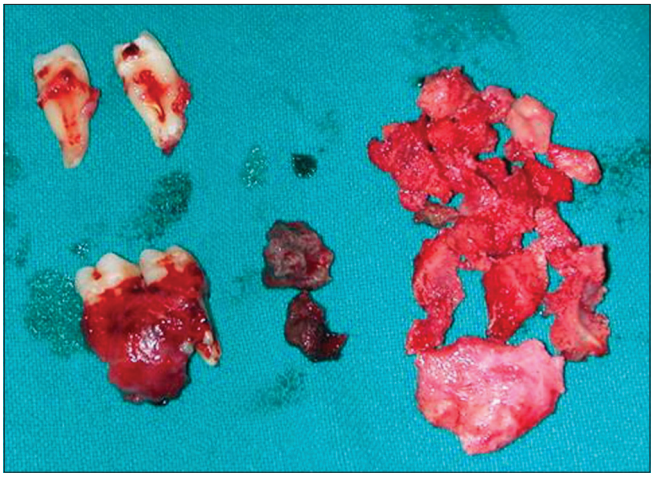

Figure 4: Pathologic specimens showing epithelial lining attached to tooth roots, pieces of sequestrum and shaved bone

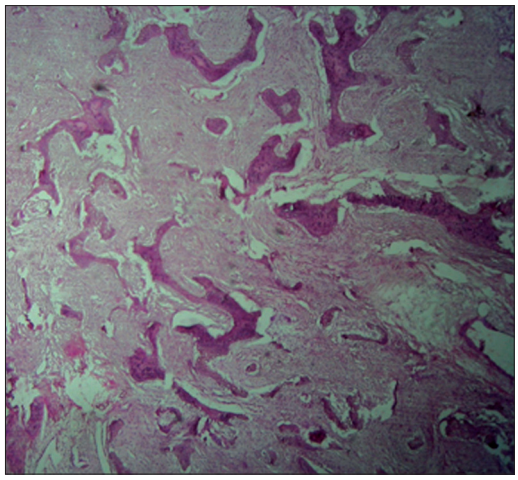

Figure 5: H and E stained 10× photomicrograph showing Chinese pattern dysplastic bone diagnostic of Fibrous Dysplasia

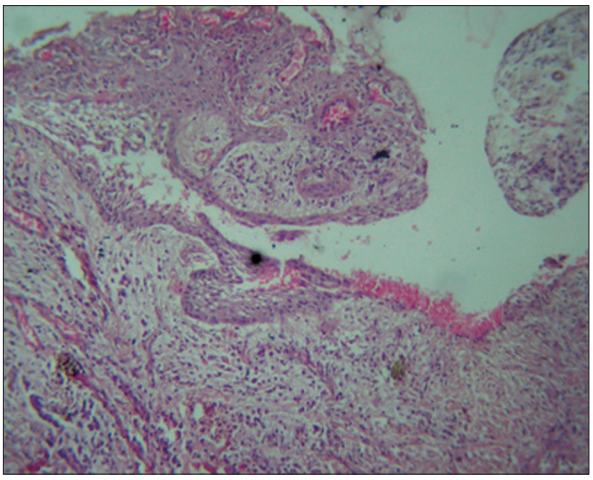

Figure 6: H and E stained 10× photomicrograph showing epithelial lining

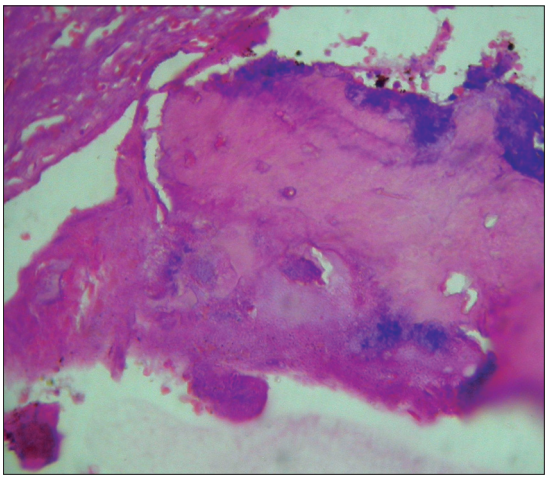

Figure 7: H and E stained 10× (zoom) photomicrograph showing avascular bone

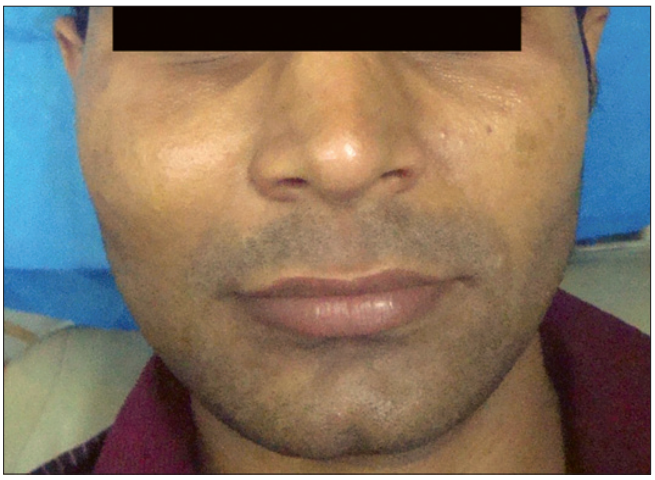

Figure 8: Post op (3 year) showing facial enlargement as a result of continued growth of fibrous dysplasia

DISCUSSION

Facial region may be affected by a form of fibrous dysplasia that is not strictly monostotic, but may be confined to a single anatomical region. These lesions affect primarily the maxilla and simultaneously cross sutures and enter into the adjacent facial bones. This type of fibrous dysplasia does not meet the precise criteria for the monostotic or polyostotic forms and has been termed craniofacial fibrous dysplasia.[11,12] In the present case, fibrous dysplasia affected the maxilla and adjacent zygomatic bone and thus may be termed as craniofacial type.

As per the current belief fibrous dysplasia results from a defect in bone maturation that begins in the embryo.[13] The classical division of fibrous dysplasia into monostotic, polyostotic and McCune Albright forms may reflect the timing of the mutation and thereby, the initial size of the mass of fibrous dysplasia precursor cells.[14] The polyostotic form may arise in foetal life whereas the monostotic form may arise postnatally. This correlates with the evidence that the monostotic form is not a precursor of the polyostotic form. Thus fibrous dysplasia may reflect a programmed field effect of abnormal osseous development in congenitally predisposed bone matrix.[15] This may account for the fusiform expansion of affected bone. Although the shape of affected maxilla appears to be more complex, reflecting its structure, the overall effect is similar to that seen in the mandible. The lesion, if large completely obliterates the maxillary sinus. The above pattern is altered if the fibrous dysplasia undergoes cystic degeneration, then the affected part may lose its anatomical shape and becomes spherical.[16] Aneurysmal bone cyst and simple bone cyst are well defined entities that sometimes occur as secondary phenomenon in many benign and malignant bone tumours.[1,17,18] Secondary cystic lesions consisting of blood filled cavities in bone that are lined by a thick layer of fibrous tissue termed as nonspecific cystic degeneration have also been reported. The development of cystic degeneration in fibrous dysplasia can pose a diagnostic and therapeutic dilemma as it may present clinically as a rapidly enlarging mass that can be alarming to both patient and physician. Sarcomatous transformation

should always be ruled out in such clinical presentations.[16,19,20] Another clinical implication of cystic degeneration, and consequentially ensuing rapid growth in a relatively quiescent fibrous dysplasia, is alteration in the management protocol necessitating early surgical intervention.[16] The affected part may lose its anatomical shape and become spherical, thus may appear more cosmetically deformed. However, this is a first case of its kind reporting an epithelial lined cyst in fibrous dysplasia occurring with chronic suppurative osteomyelitis.

Maxillary osteomyelitis occurs rarely in a healthy host and fibro osseous lesions[21] particularly fibrous dysplasia,[22] which is considered as one of the local factors that predispose to this type of infection. Chang *et al.*[23] have described a case of fibrous dysplasia with chronic osteomyelitis of mandible. Osteomyelitis complicating the fibrous dysplasia affected frontal bone subsequent to trauma has been reported in the literature.[24] Sequestrum formation has been reported in fibrous dysplasia affecting tibial bone without any clinical or pathological evidence of osteomyelitis,[25] but never in maxillofacial region. Increased predisposition may be due to vascular compromise subsequent to replacement of bony medullary cavity with immature fibrocellular tissue having relatively poor vascularity.

Presence of a carious exposed tooth in such setting may initiate chronic inflammatory response which may cause:

1. Stimulation of epithelial rests of malassez resulting into initiation of a radicular cyst.
2. Propagation of an inflammatory response which may cause the usual sequelae of abscess formation and discontinuity of cyst lining.
3. Presence of chronic foci of infection leading to osteomyelitis and sequestrum formation.
4. Persistence of chronic infection resulting into enlargement of cyst lining, granulation tissue and eventually engulfing the sequestrum.

Simultaneous occurrence of three different pathologies viz. fibrous dysplasia, epithelialised inflammatory cyst and osteomyelitis with sequestrum formation at one site is an exceptional and unique pathologic event. It is vital to sort the time frame and sequence in which these three must have occurred to understand the pathogenesis. Any one of these could have been the primary event leading to rest which means that there are three possible series of events.

1. Carious exposed tooth as the first event leading to radicular cyst →formation abscess formation → initiation of Garre's type of response with fibrous dysplastic bone.
2. Existing craniofacial fibrous dysplasia →carious exposed tooth leads to formation of radicular cyst →abscess formation and rupture of cyst lining →osteomyelitis with sequestration and engulfing by the cyst
3. Existing fibrous dysplasia → carious exposed tooth with periapical infection →osteomyelitis with sequestration and sinus tract →epithelial ingrowth encircling the sequestrum

The probability of a radicular cyst occurring as the primary event in a pre-existing fibrous dysplasia is much higher (hypothesis 2) because the cyst lining was attached to the tooth roots as noted intraoperatively and in preoperative CT scans. The reverse sequence of osteomyelitis and sequestrum formation as the primary event with its surrounding granulation getting epithelialised would lead to a cyst which will be entirely disassociated from the carious teeth. Both these hypotheses are based on the presumption that maxilla was affected by fibrous dysplasia before the tooth became carious. This is supported by the patient's history and the extent of fibrous dysplasia involving maxilla and zygoma and not just confined to alveolar region (Garre's type response). Thus, it appears to be a unique scenario in which there is occurrence of radicular cyst in the bone affected by fibrous dysplasia, and osteomyelitis develops subsequently as a consequence of altered vascularity and chronic infection. The sequestrated bone eventually gets engulfed by the disintegrated cystic lining.

Recurrence has been reported following surgical removal of non epithelial lined bone cyst occurring in fibroosseous lesions and prudence dictates similar possibility with its epithelialised counterpart. Thus follow up is advised to detect any recurrences and regrowth of fibrous dysplasia.[3,10] Rapid growth in such a dysplastic bone should raise suspicion about sarcomatous or cystic transformation thus necessitating prompt intervention.

REFERENCES

1. Martinez V, Sissons HA. Aneurysmal bone cyst. A review of 123 cases including primary lesions and those secondary to other bone pathology. Cancer 1988;61:2291-304.
2. El Deeb M, Sedano HO, Waite DE. Aneurysmal bone cyst of the jaws. Report of a case associated with fibrous dysplasia and review of the literature. Int J Oral Surg 1980;9:301-11.
3. Oliver LP. Aneurysmal bone cyst. Report of a case. Oral Surg Oral Med Oral Pathol 1973;35:67-76.
4. Diercks RL, Sauter AJ, Mallens WM. Aneurysmal bone cyst in association with fibrous dysplasia. A case report. J Bone Joint Surg Br 1986;68:144-6.
5. Buraczewski J, Dabska M. Pathogenesis of aneurysmal bone cyst. Relationship between the aneurysmal bone cyst and fibrous dysplasia of bone. Cancer 1971;28:597-604.
6. Hara H, Ohishi M, Higuchi Y. Fibrous dysplasia of the mandible associated with large solitary bone cyst. J Oral Maxillofac Surg 1990;48:88-91.
7. Saito Y, Hoshina Y, Nagamine T, Nakajima T, Suzuki M, Hayashi T. Simple bone cyst. A clinical and histopathologic study of fifteen cases. Oral Surg Oral Med Oral Pathol 1992;74:487-91.
8. Simpson AH, Creasy TS, Williamson DM, Wilson DJ, Spivey JS. Cystic degeneration of fibrous dysplasia masquerading as sarcoma. J Bone Joint Surg Br 1989;71:434-6.
9. Obwegeser HL, Freihofer HP Jr, Horejs J. Variations of fibrous dysplasia in the jaws. J Maxillofac Surg 1973;1:161-71.
10. Fisher AD. Bone cavities in fibro-osseous lesions. Br J Oral Surg 1976;14:120-7.

11. Waldron CA. Fibro-osseous lesions of the jaws. J Oral Maxillofac Surg 1993;51:828-35.
12. MacDonald-Jankowski DS. Fibro-osseous lesions of the face and jaws. Clin Radiol 2004;59:11-25.
13. Marx RE Fibro-ossoeus diseases and systemic diseases affecting bone. In: Marx RE, Stern D. Editors. Oral and Maxillofacial Pathology: A rationale for diagnosis and treatment. 1st ed. Illinois: Quintessence publishing co. inc; 2003. p. 747.
14. Chapurlat RD, Meunier PJ. Fibrous dysplasia of bone. Baillieres Best Pract Res Clin Rheumatol 2000;14:385-98.
15. Eisenberg E, Eisenbud L. Benign fibro-osseous diseases: Current concepts in historical perspective. Oral Maxillofac Clin Nor Am 1997;9:551-62.
16. Ferretti C, Coleman H, Dent M, Altini M. Cystic degeneration in fibrous dysplasia of the jaws: A case report. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1999;88:337-42.
17. Trent C, Byl FM. Aneurysmal bone cyst of the mandible. Ann Otol Rhinol Laryngol 1993;102:917-24.
18. Svensson B, Isacsson G. Benign osteoblastoma associated with an aneurysmal bone cyst of the mandibular ramus and condyle. Oral Surg Oral Med Oral Pathol 1993;76:433-6.
19. Schwartz DT, Alpert M. The malignant transformation of fibrous dysplasia. Am J Med Sci 1964;247:1-20.
20. Eversole LR, Sabes WR, Rovin S. Fibrous dysplasia: A nosologic problem in the diagnosis of fibro-osseous lesions of the jaws. J Oral Pathol 1972;1:189-220.
21. Kushner GM, Alpert B. Osteomyelitis and Osteoradionecrosis. In: Miloro M. Editor. Peterson's Principles of Oral and Maxillofacial Surgery. 2nd ed., vol. 1. London: BC Decker; 2004. p. 313.
22. Topazian RG. Osteomyelitis of the Jaws. In: Topazian RG, Goldberg MG, Hupp JR, Editors. Oral and Maxillofacial infections. 4th ed., Philadelphia: WB Saunders Co; 2002.
23. Chang CY, Wu KG, Tiu CM, Hwang B. Fibrous dysplasia of mandible with chronic osteomyelitis in a child: Report of one case. Acta Paediatr Taiwan 2002;43:354-7.
24. Williams GT, Anderson W, Bryce DP. Osteomyelitis complicating fibrous dysplasia of the skull. Arch Otolaryngol 1972;96:278-81.
25. Pratt AD, Felson B, Wiot JF, Paige M. Sequestrum formation in fibrous dysplasia. Am J Roentgenol Radium Ther Nucl Med 1969;106:162-5.

How to cite this article: Jhamb A, Mohanty S, Jhamb PA. Craniofacial fibrous dysplasia. J Oral Maxillofac Pathol 2012;16:465-9.

Source of Support: Nil. **Conflict of Interest:** None declared.