Original Article

The radiological versatility of fibrous dysplasia: An 8-year retrospective radiographic analysis in a north Indian population

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Abstract

Objective: The aim of this study was to examine the clinical and radiographic presentation of fibrous dysplasia through an 8-year retrospective study in patients who reported to the outpatient unit of the Kothiwal Dental College and Research Centre, Moradabad. **Materials and Methods:** The clinical file records and radiographs of the patients who reported to the outpatient clinic in the Department of Oral Medicine and Radiology were retrospectively reviewed for histopathologically diagnosed fibrous dysplasia. A detailed analysis of the clinical and radiographic features of the 14 cases retrieved was carried out. **Results:** Almost all the patients presented with a complaint of swelling on the side of the face. The maxilla was more frequently involved than the mandible. The most common radiographic pattern observed was the ground-glass appearance, followed by orange peel, cotton wool, sunray and thumb print appearance, which leads to a perplex differential diagnosis. **Conclusion:** Awareness of the versatile features of fibrous dysplasia evident through this study is essential in the accurate diagnosis and proper treatment planning of such lesions.

Key words: Cotton wool, fibrous dysplasia, ground glass, orange peel, thumb print

INTRODUCTION

Fibrous dysplasia is a non-neoplastic hamartomatous developmental lesion of the bone of unknown origin. It is characterized by the replacement of bone with fibro-osseous tissue, as given by Waldron in 1985.[1] There are two primary categories of the disease: Monostotic fibrous dysplasia that involves only one bone and polyostotic fibrous dysplasia, which involves several bones. A monostotic form does not progress into a polyostotic form of the disease. The Jaffe - Lichtenstein syndrome is a variant of polyostotic fibrous dysplasia with cafe'-au-lait pigmentation of the skin. A more severe form of the polyostotic type of fibrous dysplasia accompanied by endocrine disturbances of varying types in addition to skin pigmentation is called the McCune-Albright syndrome. Another category identified as the craniofacial type of fibrous dysplasia is confined to the face and jaws involving two or more bones.[2] Polyostotic fibrous dysplasia with soft tissue myxomas is called Mazabraud syndrome.[3] Fibrous dysplasia may be divided into three categories: Monostotic (74%), polyostotic (13%) and craniofacial (13%).[4] Fibrous dysplasia creates radiographic patterns that are virtually indistinguishable from other lesions affecting the bones, such as Paget's disease and cemento-osseous fibroma. Difficulty is often experienced when the lesion is detected on radiographs with negligible clinical evidence.[2] Over the past few decades, there are considerable number of case reports published on the various radiological features of fibrous dysplasia. Its radiological versatility most of the time gives a perplex diagnosis leaving it on a controversial note. Here, we report a series of 14 cases who reported to the outpatient department of the Kothiwal Dental College and Research Centre, Moradabad. All of them featured myriad radiology that gave versatile diagnosis of fibrous dysplasia confirmed by histopathological reports.

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MATERIALS AND METHODS

A review of the radiology records of the Department of Oral Medicine Diagnosis and Radiology, Kothiwal Dental College and Research Centre, Moradabad between 2005 and 2012 revealed 14 patients with fibrous dysplasia where the diagnosis had been histopathologically confirmed. Each patient's name, age, sex, clinical history, findings on examination and the provisional and differential diagnosis were reviewed. Their clinical notes and radiographs were retrospectively reviewed and analyzed. Institutional Ethical Committee approval was not required as this was a retrospective study based on file records of patients who gave informed consent prior to documentation and treatment.

Patients with inadequate histories and irrelevant radiographic data were not included in this series. Panoramic radiographs were available in 12 cases. Intraoral periapical and occlusal radiographs supplemented by skull views and conventional or three-dimensional computed tomography (CT) were performed where appropriate. All the features were entered into a proforma highlighting the radiological features. Intraoral periapical radiographs were considered for all cases at the suspected peripheries of the lesions in order to define the exact extent of the lesion. CT was performed in few cases when the borders of the lesion could not be discerned from plain radiographs, especially in the lesions affecting the anatomically complex maxilla. The radiographs were reviewed on a standard illuminated screen. The various radiographic parameters studied were the periphery of the lesion; internal structure; presence of areas of cystic degeneration; effects on adjacent structures, including displacement of inferior alveolar canal; encroachment into paranasal sinuses; expansion of the lower border of the mandible; thinning or loss of cortex; loss of lamina dura; and root resorption. The accepted radiological criteria from the literature and standard text books such as White and Pharaoh and Worth were considered.[5,6] The definition of boundaries was according to the criteria set by Slootweg and Muller.[7] A lesion was considered to be well demarcated when its radiodensity changed significantly within a distance of 1 mm when passing to the surrounding bone.

RESULTS

Patient demographics

During the 8-year period of the study, 14 cases were histopathologically diagnosed as fibrous dysplasia. The cases were in the age range of 12-62 years at presentation, with an average age of 29.6 years. An increased prevalence of the disease was seen in the second and third decades of life. Male and female patients accounted for seven cases each. The females were older than the males: The mean age of the males was 23.2 years and that of the female was 34.2 years. The chief complaint of 13 patients was an esthetically disfiguring swelling of the face, and one patient reported of swelling with pain. She also gave a previous history of surgery where cosmetic curettage was performed 10 years ago. Eight cases were classified as having monostotic lesions, five cases had craniofacial lesions and one case had a polyostotic presentation.

Anatomic location of lesions

Among the cases of fibrous dysplasia affecting the facial bones, five cases only involved the mandible and four cases only involved the maxilla. Among the five lesions involving the craniofacial complex, three were only involving the maxilla and two were involving both maxilla and mandible. They also involved the frontal, zygomatic, temporal, sphenoid, ethmoid, nasal bone and floor of the orbit. In the single case of polyostotic fibrous dysplasia apart from face, there were cafe'-au-lait pigmentations on his right hand. The unilateral presentation of fibrous dysplasia was apparent in 12 cases in the present study, with an increased predilection for the right side. Among the cases involving the maxilla, the premolar–molar area was affected in three cases; the canine premolar–molar area in three cases; and only molar area in one case. Among the cases affecting the mandible, the body and angle of the mandible was involved in three cases; the angle region was affected in one case; and the symphyseal and parasymphyseal region of the right and left sides in one case. The entire maxilla and mandible were affected unilaterally in two cases.

Radiographic analysis of the lesion

Of all the cases reviewed, four cases were investigated by CT. Only four cases, of which three cases involved the mandible and one case involved the maxilla, showed a well-defined corticated border. The other 10 had ill-defined peripheries. Nine cases presented with ground-glass appearance [Figures 1a-d, 2a-e, 3a-c and 4a-c], one with orange peel [Figure 5a and b], two with cotton wool [Figures 5c and 6a], one with sunray [Figures 5d and 6b] and one with thumb print appearance [Figure 6c]. Twelve cases were unilateral and two were bilateral. The shape of the mandibular lesions was fusiform whereas the maxillary lesions generally followed the shape of the bone. All lesions had expanded the alveolar process bucco-lingually. All four cases affecting the

mandible had caused downward displacement of the inferior border of the mandible, whereas three cases were associated with downward displacement of the inferior alveolar canal [Figures 1a and 4a] and in one case, it was indiscernible [Figure 5c]. Four lesions revealed effacement of the floor of the maxillary sinus [Figures 1d, 2d and 3c] and five cases revealed its obliteration [Figure 3a]. The case with orange peel appearance revealed a cystic lesion in the 33 and 34 tooth regions apically [Figure 5b].

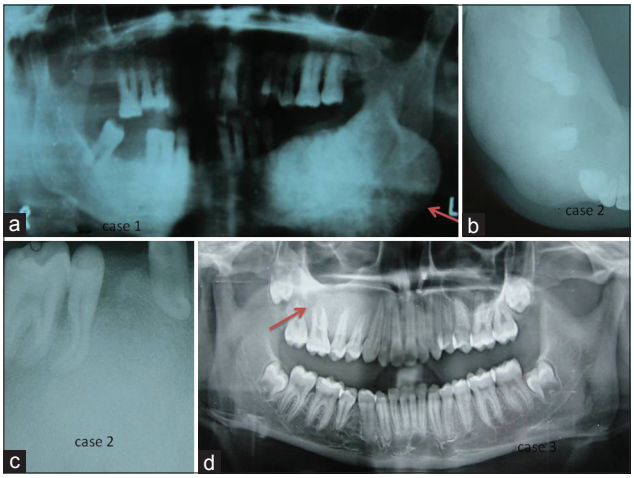

Figure 1: (a) An orthopantomogram of case 1 revealing ground-glass appearance in the left mandible and having a fusiform shape. (b) A mandibular right lateral cross-sectional occlusal radiograph of case 2 showing ground-glass appearance. (c) An intraoral periapical radiograph of case 2 showing ground-glass appearance and dilacerations of the roots. (d) An orthopantomogram of case 3 revealing ground-glass appearance in the right premolar -- molar region

DISCUSSION

This clinical series demonstrates the analysis of the various radiographic presentations of fibrous dysplasia [Table 1]. There are variations in this study compared with the earlier reports and studies on fibrous dysplasia. The average age of presentation was 29.75 years. This was the age at which the cases presented to the clinic. Two patients over the age of 50 years presented with a massive swelling, indicating that such lesions may remain asymptomatic and may stop patients from seeking dental care until there is gross disfigurement or pain is experienced. In case 1, the patient gave a history of swelling of more than 15 years duration but refused treatment because the lesion was not increasing in size. This is in accordance with Eisenberg and Eisenbud, who stated that the majority of cases burn out in early adulthood when skeletal maturity has been reached.[8] A single male patient in the third decade of life presented with the polyostotic form consistent with the findings of Windolz,[9] who revealed that the polyostotic form typically represented

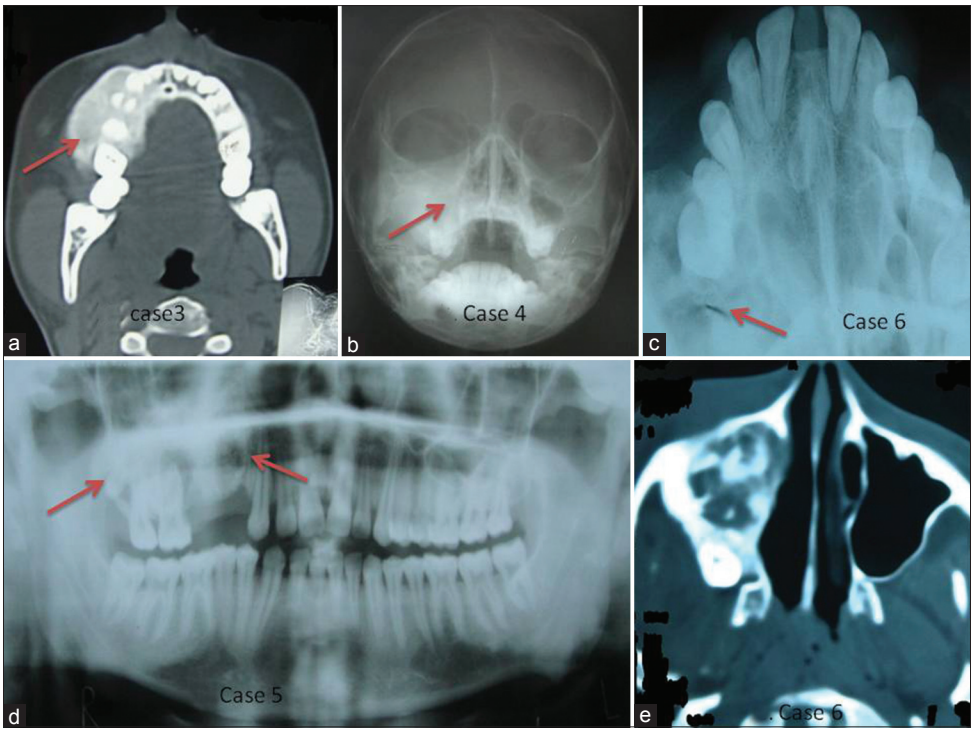

Figure 2: (a) An axial computed tomography of case 3 revealing the extensions of the lesion in the right maxilla. (b) An occipitomental view of case 4 revealing ground-glass appearance in the right maxilla. (c) Occlusal view of case 6 showing ground-glass appearance in the right maxilla. (d) Orthopantomogram of case 5 revealing ground-glass appearance in the right maxilla. (e) Coronal computed tomography of case 6 revealing a large expansile lesion in the right maxilla

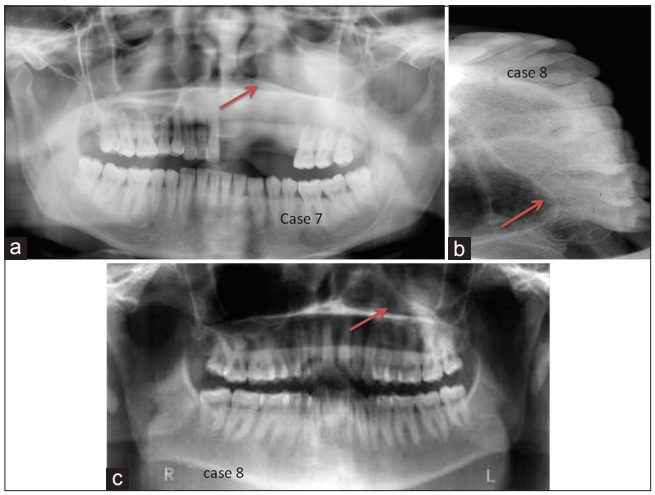

Figure 3: (a) Orthopantomogram of case 7 revealing ground-glass appearance in the left maxilla. (b) Maxillary left cross-sectional occlusal radiograph of case 8 revealing an acute angle made by the floor of the maxillary antrum. (c) Orthopantomogram of case 8 showing an acute angle of the wall of the maxillary sinus on the left side

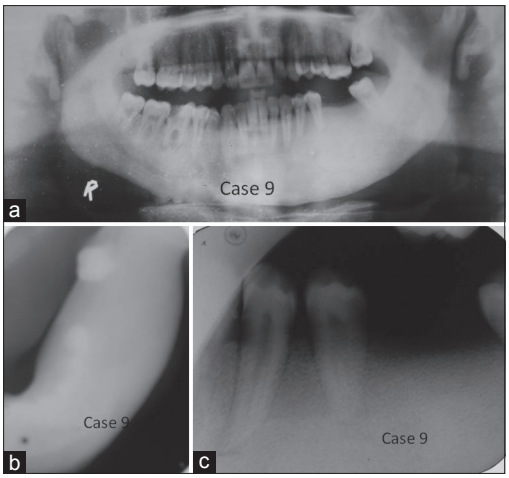

Figure 4: (a) Orthopantomogram of case 9 revealing ground-glass appearance in the left mandible. (b) Mandibular left lateral cross-sectional occlusal radiograph of case 9 revealing ground-glass appearance. (c) An intraoral periapical radiograph of case 9 revealing ground-glass appearance

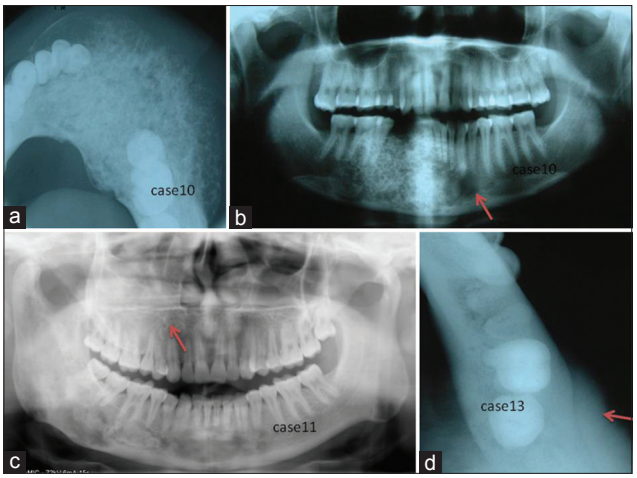

Figure 5: (a) Mandibular right lateral cross-sectional occlusal radiograph of case 10 revealing orange peel appearance. (b) Orthopantomogram of case 10 revealing orange peel appearance with a cystic variety. (c) Orthopantomogram of case 11 revealing cotton wool appearance in the mandible and maxilla involving the craniofacial complex. (d) Mandibular left lateral cross-sectional occlusal radiograph of case 13 showing sunray appearance

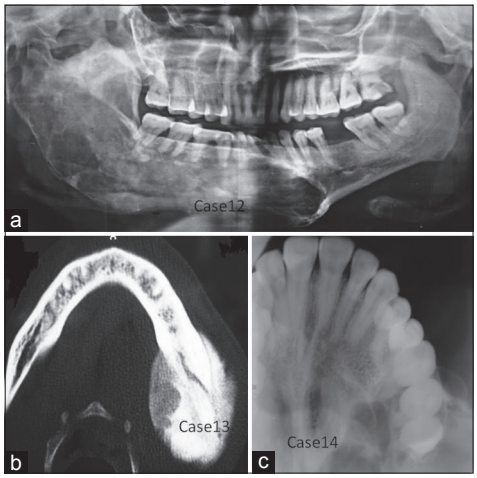

Figure 6: (a) Orthopantomogram of case 12 revealing sacromatous transformation in the mandible and maxilla involving the craniofacial complex. (b) Axial computed tomography of case 13 showing sun spicule pattern. (c) Maxillary left lateral cross-sectional occlusal radiograph of case 14 revealing thumb print appearance in the molar region

only about one-third of patients. In our study, females and males were equally affected, which was not similar with the systematic review of cases of fibrous dysplasia by MacDonald-Jankowski,[4] Yoon *et al*.[10] and Slootweg and Muller,[7] where a female predominance was noted. Like most studies on fibrous dysplasia, in our clinical series, the maxilla was more commonly affected than the mandible.[4,7,10] A predilection for the right side was observed in this study; however, it may be expected that such a predilection may just be an earlier stage of manifestation of the disease condition in monostotic patients and may vary when the sample size is larger.[4,11]

Craniofacial bones were involved in five cases, of which two cases also involved the mandible, including the case showing a polyostotic presentation. With regard to involvement of the bones of the craniofacial region, the maxilla was the most commonly involved bone, followed by the mandible, zygoma, frontal, temporal, sphenoid, ethmoid and parietal. Such a presentation is in contrast with the findings of Lustig and co-workers,[12] who identified the ethmoid as the most commonly involved bone, followed by the sphenoid, frontal, maxilla, temporal, parietal and occipital bones.

This article highlights the various radiographic presentations of fibrous dysplasia in the jaw bones.

Table 1: Summary of the clinical and radiographic features of 14 case series

Case no.	Age	Sex	Chief complaint with duration	Site of the lesion	Periphery of the lesion as seen in the radiograph	Appearance of the lesion as seen in the radiograph
1	62	Female	Swelling since 15 years	Mandible 34-37 region	Well defined	Ground glass
2	13	Male	Swelling since 4 years	Mandible 41-47 region	Ill defined	Ground glass
3	19	Female	Swelling since 5 years	Maxilla 11-18 region	Well defined	Ground glass
4	12	Female	Swelling since 3 years	Maxilla 12-16 region	Ill defined	Ground glass
5	38	Male	Swelling since 5 years	Maxilla 13-18 region	Well defined	Ground glass
6	47	Female	Swelling since 9 years	Craniofacial complex and 12-18 region	Ill defined	Ground glass
7	25	Male	Swelling since 10 years	Craniofacial complex and 21-28 region	Ill defined	Ground glass
8	20	Male	Swelling since 8 years	Craniofacial complex and 14-17 region	Ill defined	Ground glass
9	18	Male	Swelling since 6 months	Mandible 32-37 region	Ill defined	Ground glass
10	27	Female	Swelling since 7 years	Mandible 47-34 region	Ill defined	Orange peel with cystic variety
11	23	Male	Swelling since 10 years	Craniofacial complex, 12-18 region and mandible with café-au-lait pigmentation on the right hand	Ill defined	Cotton wool
12	54	Female	Swelling with pain since 10 years after surgery in the same region	Craniofacial complex and mandible	Ill defined	Cotton wool
13	26	Male	Swelling since 4 years	Mandible 37-38 region involving the ramus and angle	Ill defined	Sunray
14	19	Female	Swelling since 2 months	Maxilla 26-28 region	Well defined	Thumb print

To a certain extent, plain films help to define the lesion. CT scans are required to demarcate lesions in the anatomically complex maxilla and the skull. Periapical radiographs however do help to determine the internal structure of fibrous dysplasia accurately as it is placed closest to the jaw. Only four of the 14 cases showed well-defined borders, three of which involved the maxilla [Figures 1a and d and 2d and 6c]. The lesion may be sharply defined when a sclerotic rim is present or may be ill defined if it lacks perilesional sclerosis and may fade into the adjacent normal bone.[13] Sherman and co-workers[14] and Eversole *et al*.[15] reasoned that diffuse lesions are usually poorly defined and dysplastic in nature. The basic patterns described by Fries in 1957 are pagetoid (56%), a mixture of dense and radiolucent areas of fibrosis; sclerotic (23%), mass is homogeneously dense; and cystic (21%), a spherical or ovoid lucency surrounded by a dense boundary.[16] In this clinical series, the most common presentation was that of a mixed radiolucent–radiopaque-type pattern, of which the "ground-glass" appearance was most common. In the study by MacDonald-Jankowski and Li[17] and in the systematic review by MacDonald-Jankowski,[4] fibrous dysplasia most commonly presented radiographically as a poorly defined ovoid (fusiform) area of dysplastic bone exhibiting a ground-glass appearance. Cases 1 to 9 presented with ground-glass appearance, which gave a differential diagnosis of juvenile ossifying fibroma or cementifying fibroma [Figures 1a-d, 2a-e, 3a-c and 4a-c]. In fibrous dysplasia, the boundaries of the lesion blends in with the surrounding bone. Moreover, it has a homogeneous appearance in the maxilla. In displaces the lateral wall of the maxilla into the maxillary antrum, maintaining the outer shape of the wall[5] [Figure 2e]. The conformational diagnosis of fibrous dysplasia is based on the bone pattern that is altered around the teeth without displacement of the teeth from one specific epicenter. The shape of the bone expansion into the antrum reflects the original outer contour of the central wall, which is different from the more convex extension of a neoplasm.[18] Case 8 revealed an acute angle of the floor of the maxillary sinus wall seen in the mandibular lateral occlusal radiograph [Figure 3b and c]. This gives a conformational diagnosis for fibrous dysplasia.

Another disease that also revealed ground-glass appearance is hyperparathyroidism. In this, the overall effect is one of rarefaction but not of radiopacity. The radiographic pattern in this disease is generalized, not localized or solitary as in fibrous dysplasia. Caudill *et al*. concluded that maxillofacial lesions represent the polyostotic fibrous dysplasia component of the Albright syndrome. The endocrine abnormality could be manifested by the hyperparathyroidism.[19]

Case 10 presented with orange peel appearance with a cyst in the same anterior region. Cystic change was noted in one case in a patient who was in the third decade of life thus justifying that radiolucent lesion resembling cysts occasionally occur in mature lesions of fibrous dysplasia [Figure 4a].

Case 11 presented with cotton wool appearance [Figure 5c]. They gave a differential diagnosis of Paget's disease. In fibrous dysplasia, a section of bone is involved that is different from Paget's disease

in which the complete bone is involved, having an asymmetric enlargement. In the maxillary extension, there is encroachment of the antral wall in fibrous dysplasia whereas in Paget's disease, there is no encroachment.[1] Alsharif *et al*.[20] and Moshy *et al*.[21] reported a spectrum of radiolucent, radiopaque and "cotton wool" appearances for fibrous dysplasia and lucency, central opacification and dense opacification with sclerotic cortices for ossifying fibroma. The case 12 patient had undergone sarcomatous changes after surgery, which were confirmed by the histopathlogy report [Figure 6a]. Gon *et al*. reported a similar case of fibrous dysplasia revealing malignant transformation that occurs in less than 1% of cases, with a mean lag period of 13.5 years.[22]

Case 13 presented with sunray appearance, which gave the differential diagnosis of osteomyelitis or osteogenic sarcoma [Figures 4d and 5b]. In osteomyelitis, the new bone that enlarges the jaws is laid down by the periosteum and therefore is on the outside of the outer cortical plate, whereas in fibrous dysplasia new bone is manufactured on the outside of the mandible.[18] Prapayasatok *et al*. reported a case of fibrous dysplasia that exhibited an unusual sunray appearance. They concluded that "sunray" or "sunburst," or sometimes "spicule" are used for periosteal new bone formation in which new bone is formed perpendicular to the cortex resulting in multiple lines like sunrays. This event could occur when the periosteum is rapidly stripped from the cortex.[23]

Cases 10 and 12 had bilateral appearance, which presents a differential diagnosis of cherubism. Cherubism occurs in early childhood between the ages of 2 and 6 years. In our case, both the patients were in their third and fifth decades of life.[5] Jain *et al*. reported that fibrous dysplasia is not usually familial, presents slightly later between 10 and 30 years of age, does not show the typical "cherubic" look and the lesions do not have a tendency to regress after puberty. In addition, the multiloculated, ground-glass lesions of fibrous dysplasia are rarely as symmetrical as the cherubic lesions.[24]

Case 14 revealed a typical thumbprint appearance in the posterior maxilla [Figure 6c]. It is similar to the report presented by Singer *et al*., revealing a similar kind of appearance in the mandible.[25]

CONCLUSION

After presenting symptoms because of the myriad radiological appearances of fibrous dysplasia, it is a challenge for the diagnostician to make an accurate diagnosis. A dentist should be aware of the versatile radiology of this dynamic disease and should keep in mind each appearance, giving a keen observation, such that an exact diagnosis can be made.

REFERENCES

1. Langlais RP, Langland OE, Nortje CJ. Diagnostic Imaging of the Jaws. Malvern: Williams and Wilkins; 1995. p. 576-82.
2. Nityasri V, Haris PS, Bose T, Balan A. Fibrous dysplasia: A 13-year retrospective radiographic analysis in a south Indian population. Dentomaxillofac Radiol 2011;40:282-9.
3. Vennila P, Chandrasekar T, Menon SS. A variant of monostotic craniofacial type of fibrous dysplasia: A rare case report. Indian J of Multidisciplinary Dentistry 2012;2:460-4.
4. MacDonald-Jankowski D. Fibrous dysplasia in the jaws of a Hong-Kong population: Radiographic presentation and systematic review. Dentomaxillofac Radiol 1999;28:195-202.
5. White SC, Pharoah M. Oral Radiology Principles and Interpretation. 5th ed. St Louis: CV Mosby; 2004. p. 485-91.
6. Worth HM. Principles practice oral radiologic interpretation. Chicago: Year Book Medical Publishers; 1963. p.607.
7. Slootweg PJ, Müller H. Differential diagnosis of fibro-osseous jaw lesions. A histological investigation on 30 cases. J Craniomaxillofac Surg 1990;18:210-4.
8. Eisenberg E, Eisenbud L. Benign fibro-osseous diseases: Current concepts in historical perspective. Oral Maxillofac Clin Nor Am 1997;9:551-62.
9. Windholz F. Cranial manifestations of fibrous dysplasia of bone; their relation to leontiasis ossea and to simplebone cysts of the vault. Am J Roentgenol Radium Ther 1947;58:51-63.
10. Yoon JH, Kim J, Lee CK, Choi IJ. Clinical and histopathological study of fibro-osseous lesions of the jaws. Yonsei Med J 1989;30:133-43.
11. Waldron CA, Giansati JS. Benign fibro-osseus lesions of the jaws: A clinical-radiologic-histologic review of sixty-five cases. Oral Surg Oral Med Oral Pathol 1973;35:190-201.
12. Lustig LR, Holliday MJ, McCarthy EF, Nager GT. Fibrous dysplasia involving the skull base and temporal bone. Arch Otolaryngol Head Neck Surg 2001;127:1239-47.
13. Bertoni F, Fernando Arias L, Alberghini M, Bacchini P. Fibrous dysplasia with degenerative atypia: A benign lesion potentially mistaken for sarcoma. Arch Pathol Lab Med 2004;128:794-6.
14. Sherman RS, Glauser OJ. Radiological identfication of fibrous dysplasia of the jaws. Radiology 1958;71:553-8.
15. 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.
16. Megerian CA, Sofferman RA, McKenna MJ, Eavey RD, Nadol JB Jr. Fibrous dysplasia of the temporal bone: Ten new cases demonstrating the spectrum of otologic sequelae. Am J Otol 1995;16:408-19.
17. MacDonald-Jankowski DS, Li TK. Fibrous dysplasia in a Hong Kong community: The clinical and radiological features and outcomes of treatment. Dentomaxillofac Radiol 2009;38:63-72.
18. Wood NK, Goaz PW. Differential Diagnosis of Oral and Maxillofacial Lesions. 5th ed. St Louis, Missouri: Mosby; 1997. p. 363-5.
19. Caudill R, Saltzman D, Gaum S, Granite E. Possible relationship of primary hyperparathyroidism and fibrous dysplasia: Report of case. J Oral Surg 1977;35:483-90.

20. Alsharif MJ, Sun ZJ, Chen XM, Wang SP, Zhao YF. Benign fibro-osseous lesions of the jaws: A study of 127 Chinese patients and review of the literature. Int J Surg Pathol 2009;17:122-34.
21. Moshy J, Dimba E, Ocholla T, Chindia M. Characteristic radiological and histological patterns of fibrous dysplasia and ossifying fibroma of the jaws at University of Nairobi Dental Teaching Hospital. Sci Res 2012;3:189-93.
22. Gon S, Majumdar B, Bhattacharyya A, Bhattacharya RN. Fibrous dysplasia: Rapid malignant transformation into osteogenic sarcoma: A rare occurance. J Pathol Nepal 2012;2:335-7.
23. Prapayasatok S, Iamaroon A, Miles DA, Kumchai T. A rare, radiographic 'sunray' appearance in fibrous dysplasia. Dentomaxillofac Radiol 2000;29:245-8.
24. Jain V, Sharma R. Radiographic, CT and MRI features of Cherubism. Pediatr Radiol 2006;36:1099-104.
25. Singer SR, Mupparapu M, Rinaggio J. Clinical and radiographic features of chronic monostotic fibrous dysplasia of the mandible. J Can Dent Assoc 2004;70:548-52.

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