

FIBRO-OSSEOUS LESIONS OF THE MAXILLO FACIAL REGION – A CASE SERIES WITH REVIEW

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ABSTRACT:

fibro-osseous lesions of the maxillofacial region are a distinct group of pathological condition in which all variations demonstrate replacement of normal bone by a fibrous connective-tissue matrix within which varying amounts or combinations of osteoid & mature bone and, in some instances, cementing like tissue are deposited. All of them have similar clinical, radiologic and histologic features. The diagnosis of these becomes mandatory because of the different treatment modalities for each of these lesions. Here in we have reported three cases of Fibro-osseous lesions in the maxillofacial region with review of literature.

Keywords: *Fibrous dysplasia, Periapical cemental dysplasia, Cemento ossifying fibroma, Cotton wool appearance, Chine's letter appearance*

INTRODUCTION

Fibro-osseous lesions (FOL) of the jaws comprise a controversial group of pathologic conditions with comparable clinical, radiological and histological features which makes diagnosis tricky. The etiology of most lesions is unknown, however some are thought to be neoplastic and others due to metabolic disorder¹. In all variants of FOL there is replacement of normal bone by fibrous tissue containing a newly formed mineralized product that may be osteoid or cementum like calcification². The lesions included under this broad category are Fibrous dysplasia (FD), Periapical cemental dysplasia, Focal and Florid cemento osseous dysplasia (FLCOD) and Cemento ossifying fibroma(OF). Fibrosa cystica, Central giant cell granuloma and Aneurismal bone cyst were also considered under FOL as they demonstrate similar microscopic feature³.

Fibro-osseous lesions have similar histologic features and their appearance does not predict the rate of growth or prognosis. These lesion exhibit a wide range of clinical and biologic behaviour which determines the management of these lesions⁴.

The aim of this article is to present a series of 3 cases of FOL with review of literature. Florid cement osseous dysplasia (FLCOD), Fibrous dysplasia(FD) and Ossifying fibroma(OF) which were evaluated according to the clinical, radiological and histopathologic findings to provide the best treatment modality for each.

CASE REPORT

CASE 1:

A 14 year old female patient reported to the OPD of VYDEHI Dental College, Bangalore with an asymptomatic swelling on the left middle half of the face. The swelling started 2 years back which gradually grew to the present size. Extra oral physical examination revealed a single diffuse swelling on the left middle third of the face, super inferiorly from the infraorbital rim to the ala tragus line. Anteroposteriorly it extended from the ala of the nose to 4cm in front of the tragus of the ear. It was nontender, bony hard in consistency with ill-defined borders (fig. 1, 2). Intraorally the swelling extended from the distal aspect of the left maxillary canine to the distal aspect of the left maxillary second molar anteroposteriorly. Inferosuperoly it extended from the free gingival margin to the mucobuccal fold. Healing socket seen with respect to left first molar. It was a non tender swelling which was bony hard in consistency with illdefined borders (fig3). Intraoral periapical radiograph of the left molar region revealed a finger print pattern of the tabacular bone. The laminadura of the teeth involved in the lesion were absent (fig4). Occlusal radiograph shows buccal cortical plate expansion (fig5). OPG (orthopantomogram) reveals a radioopaque lesion on the left maxillary region which extends from the distal aspect of the canine to the distal aspect of the second molar. The border of the lesion are ill defined the surrounding bone shows normal architecture (fig. 6). PNS view reveals involvement of the floor of the maxillary sinus of the left side (fig. 7). Histopathology revealed immature trabaculae without osteoblastic rims in less cellular fibrous stroma. The

trabeculae are curvilinear. The lesional bone is continuous with the normal compact lamellated bone at periphery, spheroidal calcifications are also seen the above features are suggestive of fibrous dysplasia of the left maxilla (fig. 8,9). Surgical recontouring was done. Patient was kept on follow-up for 6 months no recurrence was seen



Fig.1, 2: swelling seen on the left middle half of the face which extends from the inferior orbital margin to the ala tragus line



Fig. 3: buccal cortical plate expansion seen extending from 1st premolar to 3rd molar, healing socket of 1st molar seen.



Fig. 4: IOPAR shows ground glass appearance with involvement of the laminadura.



Fig. 5: Occlusal view confirms buccal cortical plate expansion.



Fig. 6: OPG shows a radiopaque lesion in the left maxilla extending from the distal aspect of left canine to the left 3rd molar region



Fig. 7: PNS shows involvement of the maxilla with no involvement of the sinus

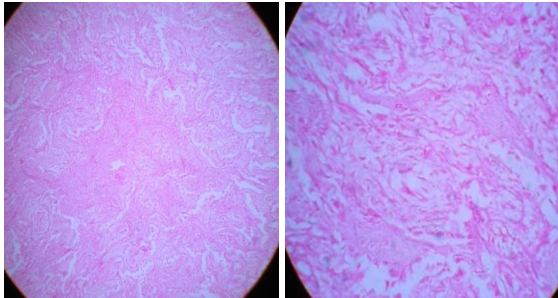


Fig. 8, 9: irregular shaped trabeculae seen in a cellular fibrous stroma

CASE 2:

A 3 year old female patient visited with a complaint of an asymptomatic swelling on the right side of the face for the past 1 year, which had gradually grown to the present size. Physical examination revealed, a single diffuse extra oral swelling measured about 3X4 cm which extended super inferiorly from the ala tragus line to the lower border of the mandible. Anteroposteriorly from the angle of the mouth to 2cm in front of the angle of the mandible. The swelling was non tender and hard in consistency with well-defined borders (fig. 10, 11). Intraorally swelling was seen in the right lower buccal vestibule. It measured about 2X2cm extending anteroposteriorly from distal aspect of right mandibular first molar to the retomolar area. Obliteration of the buccal sulcus was seen. It was hard in consistency and non tender (fig12). OPG revealed a well defined radioopaque-lucent lesion in the right body of the mandible which extended from the distal aspect of right deciduous mandibular 1st molar up to the ascending ramus. The periphery of the lesion is well defined. The right permanent 1st molar was displaced to the ramus area. Inferiorly it extended to the lower border of the mandible with involvement of the same. Resorption of the mesial and distal root of right deciduous 2nd molar is seen. Tooth bud of right 2nd premolar displaced mesially below tooth bud of right canine. The morphology of the tooth bud of right 1st premolar is lost (fig14). Histopathology revealed highly cellular fibrovascular connective tissue exhibiting numerous areas of immature cellular trabeculae of bone. The trabeculae of bone also demonstrates osteoblastic rim (fig. 15, 16). These features are suggestive of Ossifying fibroma. A final diagnosis of juvenile Ossifying fibroma was considered based on the history clinical, radiology and histopathological presentation. Excision of the lesion was done enmass. Patient was kept on follow up for 3 months no recurrence was noted



Fig. 10, 11: swelling seen in the lower 3rd of the face extending 1cm below the ala tragus line to the inferior boarder of the mandible

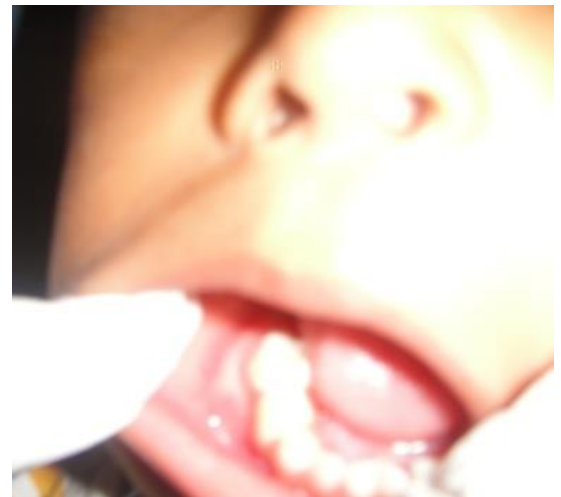


Fig. 12: obliteration of the mucobuccal fold which extends from the left canine to the first molar.

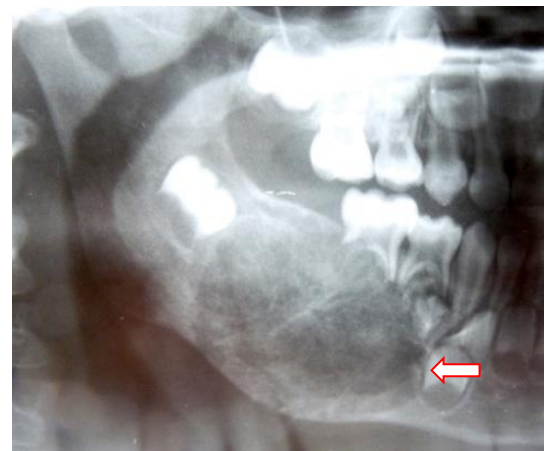


Fig: 13: A mixed radiolucency seen in the left body of the mandible which extends from the distal root of the first deciduous molar to the ascending ramus. The periphery is well corticated.

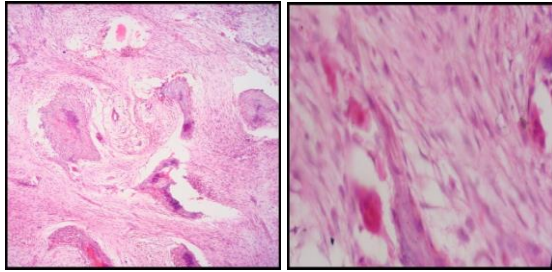


Fig. 14, 15



Fig. 16, 17: well circumscribed swelling seen in the middle half of the face.

CASE 3:

A 16 year old female patient reported with a complaint of swelling in the right side of the face since 9 years which had grown gradually. She also complained of a swelling in the left side of the face for the past 9 months. Swelling on the right middle half of the face measured about 7X6cm. Its mesiodistal extent was from lateral wall of the nose with obliteration of the nasolabial fold to 3cm in front of the tragus. Superoinferiorly it extended from the inferior orbital margin to 1cm below the ala tragus line. It was bony hard in consistency and nontender (fig17,18). Intraorally the swelling extended anteroposteriorly from the distal aspect of right lateral incisor involving the maxillary tuberosity and medially up to the mid line of the palate; laterally there was obliteration of the mucobuccal fold. Swelling was bony hard in consistency and nontender. The teeth involved in the lesion had spacing and the 2nd premolar was rotated (fig19). The swelling on the left lower half of the face extended from the symphises to 3cm in front of the angle of the mandible, it was firm in consistency with areas of softening. Intraorally the anteroposteriorly extent was from left lower canine to left 1st molar. There was missing left mandibular 2nd premolar, retained deciduous mandibular 2nd molar which was decayed (fig20). OPG showed a radioopaque lesion (cottonwool appearance) on the left maxilla with involvement of the sinus. Impacted left maxillary 2nd premolar was seen root apices of retained maxillary 2nd molar shows radioopacity with irregular borders. Ill defined radiolucency seen in the periapical region of left mandibular 2nd deciduous molar which extends

up to the inferior boarder, a breach of the lower boarder indicating fracture. Radioopacity is seen in this radiolucency which is attached to the mesial and distal root apices of left 2nd deciduous molar. Well defined radiolucency is seen in the periapical region of right mandibular first molar the boarders are not corticated (fig21). PNS also reveals involvement of the right maxillary sinus that extends up to the inferior orbital rim (fig22). Histopathology of the maxillary right lesion shows a connective tissue stroma which appears to contain trabeculae of immature bone in a moderately cellular fibrous connective tissue. Ares of basophilic globular material suggestive of cemental masses seen; these masses are seen fused to each other and with the trabaculae. No connective tissue stroma separating from the peripheral vital bone. The above features are suggestive of cemento osseous dysplasia. The maxillary left lesion also showed similar histopathologic features. The mandibular right lesion shows areas resembling cementum like masses and trabeculae of immature bone in a moderately cellular fibrous connective tissue. The connective tissue also contains a cystic lining epithelium which was non keratinised with chronic inflammatory cells. The histopathologic diagnosis of the left mandibular lesion was cemento-osseous dysplasia in association with a dentigerous cyst. A final diagnosis of florid cemento-osseous dysplasia was considered. The speciality of this is case was an occurrence of dentigerous cyst with cemento-osseous dysplasia. The patient was treated by contouring the right lesion with removal of the impacted maxillary left 2nd premolar and lower left 2nd premolar was done. Enucleation of the dentigerous cyst was performed. The patient is kept under observation for the lesion on the right side.

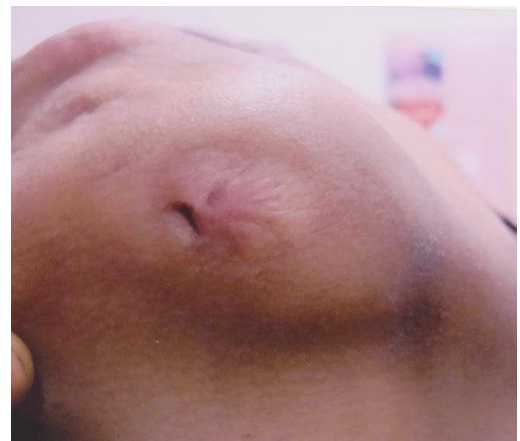


Fig. 18: healed sinus seen below the Inferior boarder of the mandible



Fig. 19: buccal and palatal expansion with rotation of 1st and 2nd premolars

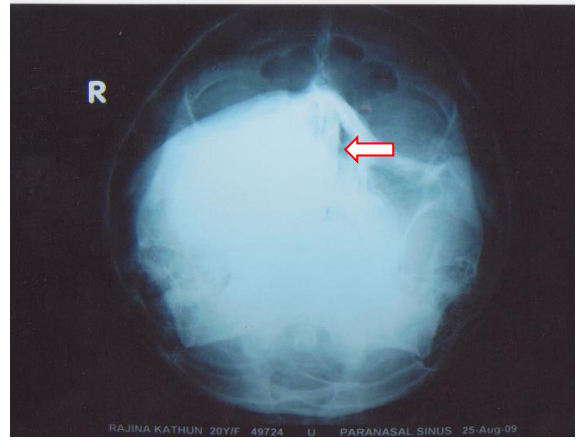


Fig 22: Radioopacity seen in the right maxillary region with involvement of the sinus



Fig. 20: obliteration of the mucobuccal fold from the cane to the 1st molar.

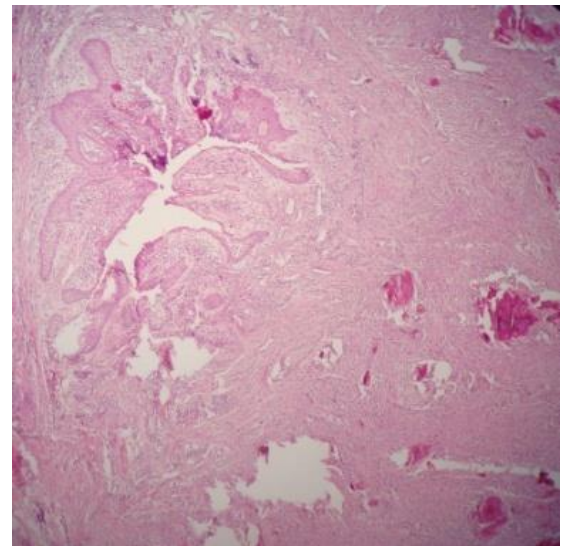


Fig 23: Shows fibrous connective tissue with areas of calcification

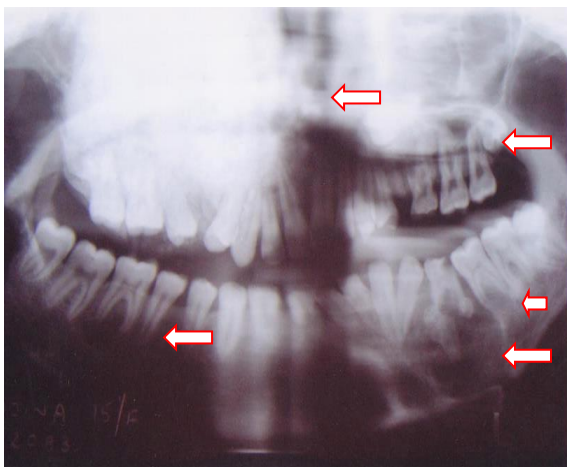


Fig 21: Cottonwool Radioopacity seen in the right maxilla with involvement of the sinus,

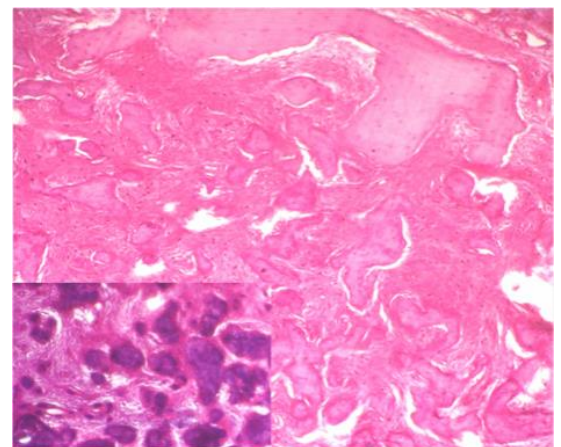


Fig 24: cementum like calcification seen

DISCUSSION

The 1930s appears to have been a time of great interest in fibro-osseous disease. A breakthrough came with the understanding of hyperparathyroidism³.

FOL of the craniofacial region are a behaviourally diversified array of disease that shares a common microscopic feature. The clinical features and imaging characteristic take a pre-eminent role in the definitive diagnosis of these lesions

Since 1930s numerous classification have evolved for the better categorization and understanding of the FOL. Complication arises with lesions like Browns tumour, CGCG (central giant cell granuloma), Aneurismal bone cyst and Pagets disease which share microscopic feature similar to FOL, however according to the

WHO 2005 classifies FOL⁵

1. Fibrous dysplasia
 - monostotic FD
 - polyostotic FD
 - craniofacial FD
2. Osseous Dysplasia
 - Periapical OD
 - Focal OD
 - Florid OD
 - . Familial Gigantiform cementoma
3. Ossifying Fibroma
 - Conventional OF
 - Juvenile trabaculæ OF
 - Juvenile psammomatoid OF
4. Central giant cell granuloma
5. Cherubism
6. Aneurismal bone cyst
7. Solitary bone cyst

We see that lesions like Pagets and Browns tumour area not included in this classification. Here in we are discussing the etiology, clinical feature, radiology, histopathology and treatment options of FD, OF and FLCOD (florid cement Osseous Dysplasia) so as to compare them.

More than 70% of FOLs arise in the head and neck region⁵⁻⁸. It is not easy to diagnose and manage the FOL in the mandible or maxilla because it poses more of a cosmetic problems and some time may lead to serious complications.

Clinical features and etiology of FOL

FD (Fibrous dysplasia) is a nonodontogenic aggressive lesion. It may affect a single bone, the monoostotic form. This is about 80% of all cases of FD. In poly ostotic form multiple bones are affected and the third variant is the McCune-Albrights

syndrome in which FD is associated with endocrinal disturbances and irregular café-au-late spots. This classification may reflect the time of mutation⁹.

In 1927 Montgomery first used the term of (Ossifying fibroma) a clinicoradiological term. Sherman and Sternberg in 1948 published a detailed description of the clinical, radiological and histological characteristics of OF¹⁰

Cemento-osseous dysplasias (COD) is probably the most common fibro-osseous lesion encountered in clinical practice. Because cemento-osseous dysplasias arise in close proximity to the periodontal ligament and exhibits histological similarities with the structures, some investigators suggest the lesion to be of periodontal origin. Others believe that it can be a defect in the extraligament bone remodelling triggered by local factors or underlying hormonal imbalance¹¹. The second edition of WHO classification in 1992 recognizes three separate COD entities, "Periapical cemental dysplasia" (PCD), "florid COD" (FCOD)^{12,13} and "other COD". According to Waldron, "they appear to represent only variants of the same disease process"¹².

Though all the FOL look similar posing difficulty in diagnosis the etiology of each is different. FD is thought to be due to a mutation in the GNAS gene^{11,14,15} whereas the exact cause of OF is not known. Eversawyer had reported identical chromosomal breakpoints in 3 of his cases of Juvenile ossifying¹⁶. F.J. Pimenta in their study for the first time showed mutation of HRPT2 gene mutation in OF and suggested that OF may arise due to haploinsufficiency of the HRPT2 gene¹⁶. FLCOD may be familial with an autosomal dominant inheritance pattern, only few such cases in literature have been reported¹⁷. Our cases also did not show any familial inheritance. FD has a female predilection with occurrence of the lesion commonly in the maxilla. The JOF variant has a more mandibular predilection¹⁸. FLCOD may be seen involving all 4 quadrants^{14,15}. The occurrence of these disease is varied, FD is seen in the first and second decades of life whereas OF is seen in the third and fourth decades of life with the juvenile variant seen more commonly between the 5 and 15 years of age. Cases have been reported in 6 months old children¹⁴, whereas FLCOD is common in middle aged black women¹⁹. Teeth in the region of FOL are usually vital. In FD there is tipping and displacement of teeth seen as it is a progressive lesion^{11,14}. Most FOL show a facial deformity which is usually asymptomatic. Some time FLOCD can present with only dull pain. In FLOCD chronic osteomyelitis can be seen where the patient may present with pain and paresthesia^{4,5}.

Radiographic findings in FOL

Radiology plays an important role in the diagnosis of FOL. FOL present as a mixed radiolucent radioopaque lesion on a radiograph. However each of these lesions have few distinct diagnostic radiographic features which compliment diagnosis. The radiographic features of FD may have an array of presentation which includes multilocular appearance, ground glass, peau-d orange appearance, cottonwool appearance. The swirling pattern of the abnormal trabecular bone presents as fingerprint appearance which is due to abnormal trabecular pattern^{21,22}. The periphery of the lesion in FD is ill defined as the lesion gradually blends with the normal bone this feature is diagnostic as it can be used to differentiate between OF. The laminadura of the teeth in the lesion is absent and displacement of inferior alveolar nerve in a superior direction is seen in mandibular lesions. In the maxilla it can displace the antrum.

OF also have a mixed radiolucent opaque appearance the internal pattern may resemble that of FD. When cementum is present an amorphous radiopacity can be seen. In contrast to FD, OF has a distinct border which separates the lesion from the normal bone. In contrast to FD the inferior alveolar canal is displaced in an inferior direction. The outer cortical plate remains intact although it is displaced and thinned out. CT images of FD on bone window can display the range of opacification observed on plain radiographs. OF exhibits thin intact cortex¹⁰ which is absent in FD. Signal intensity on T1 and T2 weighted magnetic resonance imaging (MRI) is dependent on factors such as amount of bone trabeculae and the bone cellularity²³. In some cases FD and OF were misdiagnosed as meningioma on MRI²⁴. MRI can be used to show neurovascular and ocular involvement²⁵. The FLCOD lesions appear as multiple sclerotic masses located in 2 or more quadrants usually in the tooth bearing region. They are often confined to the alveolar bone¹³. Normally epicentre is apical to the teeth. The periphery of the lesions is well defined. Rarely a simple bone cyst forms in these lesions.

Histopathology of FOL

The histopathology of all FOL is very similar. All lesions are composed of a hypercellular fibrous element and an osseous element both of which exhibit a wide spectrum of variations. FD shows irregular shaped trabeculae of immature bone in a cellular loosely arranged fibrous stroma. The trabeculae are not connected and assume curvilinear shapes resembling Chinese script. The trabeculae are considered to arise from metaplasia and not from osteoblasts. The lesions directly fuse with normal bone without any demarcation. In OF the fibrous tissue exhibits varying degree of cellularity and

contains mineralized material. The hard tissue may be basophilic and spherical resembling cementum. The trabeculae vary in size demonstrating woven and lamellar pattern. Peripheral osteoblastic rim is seen¹¹ which is absent in FD. FLCOD lesions consist of fragments of cellular metaplasia tissue composed of spindle shaped fibroblasts with neovascular small blood vessels within the fibrous connective tissue. Background is a mixture of woven bone, lamellar bone and cementum like particles. With maturation the bone trabeculae become thick curvilinear and resemble ginger roots.

Treatment of FOL

Some symptomless FOL require no surgery. So the radiological and clinical diagnosis is important to obviate the need of unnecessary surgical treatment. Exaggerated growth of FD could be stimulated by surgery in young patients so treatment is deferred until stabilization of the disease process. Cosmetic and function recontouring may be needed in large deforming lesions. The other modes include conservative enucleation, curettage or radical surgery, and segmental resection of an extensive lesion with autogenous bone graft. However, Slootweg and Muller reported that there was no difference in the outcome between patients treated in a more limited way and those treated by major surgery²⁶. Malignant transformation is reported with radiotherapy. OF needs complete enucleation or resection of the lesion. In JOF rapid enlargement is seen and needs wide resection to prevent recurrence. No malignant transformation is reported in OF cases till date. FLCOD treated before sclerosis leads to regrowth of the lesion. Extensive resection and recontouring of facial skeleton and associated soft tissue is recommended²⁷. Prognosis of FOL is good, provided that it is diagnosed correctly and with appropriate treatment plan.

CONCLUSION

FOL of the craniofacial bones are a behaviourally diversified array of diseases that share common microscopic features. Thereby the clinical presentation and imaging characteristics are of paramount importance in the diagnosis of the diseases. It becomes important that the oral clinician, radiologist, pathologist and surgeon should integrate all information to arrive at a correct diagnosis for appropriate disease management. Further research in the field of imaging is needed.

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