Case Report

DOI: 10.7860/JCDR/2015/11192.5417

Dentistry Section

# Giant Central Ossifying Fibroma of the Maxilla Presenting with a Pus Discharging Intra-Oral Sinus

SATYA RANJAN MISRA[1], ANJALI SAIGAL[2], VARUN RASTOGI[3], SMITA R. PRIYADARSHINI[4], ABHISHEK RANJAN PATI[5]

## ABSTRACT

Central ossifying fibroma (COF) is a rare benign fibro-osseous neoplasm which has a predilection for mandible and is encountered in middle aged women. It arises from mesenchymal blast cells of the periodontal ligament, and with a potential to form fibrous tissue, cementum and bone. It is a well circumscribed lesion mainly comprising of fibrous tissue with varying amounts of bone or cementum or osteo-cementum like tissue. We present an uncommon case of COF of the maxilla in a 35-year-old male who presented with a pus discharging sinus in the maxilla. Due to the bizarre size of the lesion we have termed as 'giant' COF. There is extensive facial asymmetry on the right side with obliteration of the maxillary antrum, deviation of the nose and upward displacement of the orbit.

**Keywords:** Benign fibro-osseous lesion, Huge tumour, Non-odontogenic neoplasm, Radiopaque-radiolucent lesion, Secondary infection

## CASE REPORT

A 35-year-old male patient reported to the dental hospital with the complaint of pain and pus discharge in the right maxillary molar region for two weeks.

History revealed that the patient noticed a small swelling in the right side of the face eight years back which had gradually grown to attain a huge size, on the face as well as intra-orally. There was no pain, tenderness or any discharge associated with the swelling but 15 d back the patient developed pain in the right maxillary molar teeth with pus discharge from the intra-oral swelling which did not subside on taking medications. He also had nasal stuffiness and restricted mouth opening for some months.

On clinical examination a diffuse swelling was seen in the right side of the face, ovoid in shape extending from the right inferior orbital margin to the angle of the mouth supero-inferiorly and from the bridge of the nose up to 3cm from the tragus of the ear antero-posteriorrly, about9x5cm in size, bony hard in consistency and non-tender on palpation [Table/Fig-1,2a,b]. Due to the swelling there was obliteration of the nasolabial sulcus and displacement of the bridge of the nose and the angle of the mouth. Mouth opening was restricted; the inter-incisal distance being 32mm. Intra-orally a large swelling was seen extending from the canine to second molar region, obliterating the buccal sulcus and with expansion of the buccal cortex [Table/Fig-3]. There was the presence of a pus discharging sinus in the periapical region of maxillary first premolar with flaring of the buccal root, severe gingival recession, mobility and tenderness. Correlating the history of a long standing swelling, presence of a bony swelling with considerable expansion and an intra-oral pus discharging sinus led to provisional diagnosis of a benign bony lesion with secondary infection.

### Clinical Differential Diagnosis

1. Craniofacial fibrous dysplasia
2. Infected dentigerous cyst
3. Adenomatoidodontogenic tumour
4. Central Ossifying Fibroma

### Investigations

Periapical radiograph of maxillary premolar-molar region showed indistinct trabeculae with patchy radio-opacities.

Maxillary right lateral occlusal radiograph revealed bicortical expansion with cotton wool appearance [Table/Fig-4].

Postero-anterior (PA) view of the skull, Water's view of skull [Table/Fig-5a,b] and Panoramic radiograph [Table/Fig-6] revealed a localized, well circumscribed mixed radio-opaque-radiolucent lesion in the right maxilla obscuring the antrum and displacing the orbital floor.

Computed Tomography (CT) scan of the maxillofacial region revealed a well-defined, heterodense, space occupying, and expansile lesion in the right maxilla, with bicortical expansion and perforation buccally. The lesion had displaced the floor of the orbit and the nasal cavity, entirely filling up the right maxillary antrum [Table/Fig-7- 9].

### Radiologic Differential Diagnosis

1. Craniofacial fibrous dysplasia
2. Calcifying odontogenic cyst
3. Calcifying epithelial odontogenic tumour
4. Central ossifying fibroma
5. Osteosarcoma

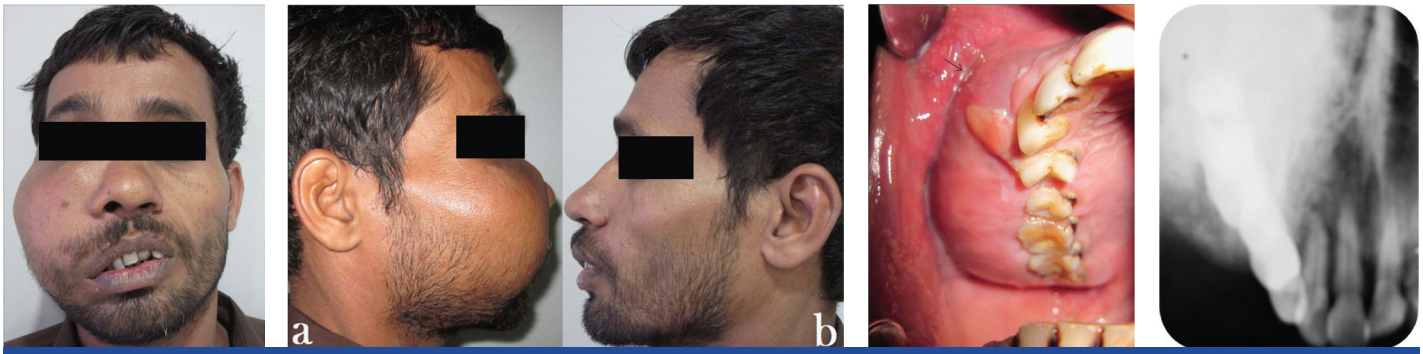
The differential diagnosis was based on radiologic findings. Presence of cotton wool appearance filling the antrum unilaterally is a feature of fibrous dysplasia, while well-defined expansile mixed radio-opaque-radiolucent appearance is a finding in calcifying odontogenic cyst, calcifying epithelial odontogenic tumour, central ossifying fibroma and osteosarcoma.

Incisional Biopsy: showed the presence of a cellular connective tissue stroma spindle and stellate shaped cells and numerous areas of calcification resembling bone along with osteoblastic rimming and cementum like areas. There was evidence of moderate vascularity. [Table/Fig-10].

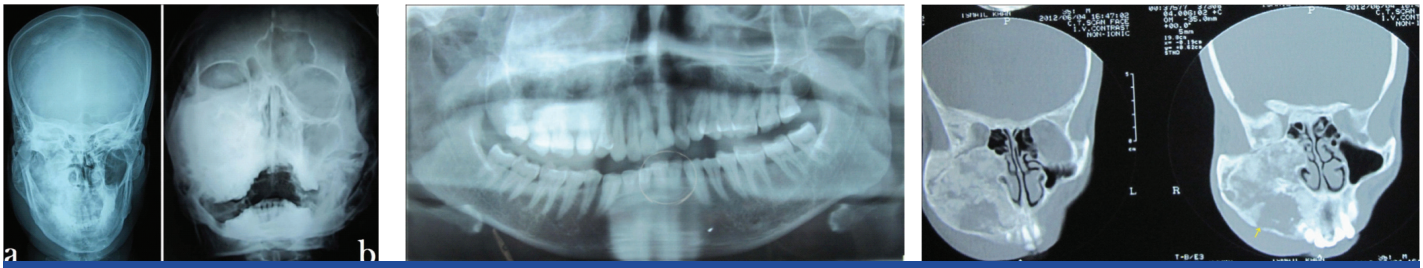
A final diagnosis of COF was made but the patient denied treatment due to financial constraints and could not be followed-up.

## DISCUSSION

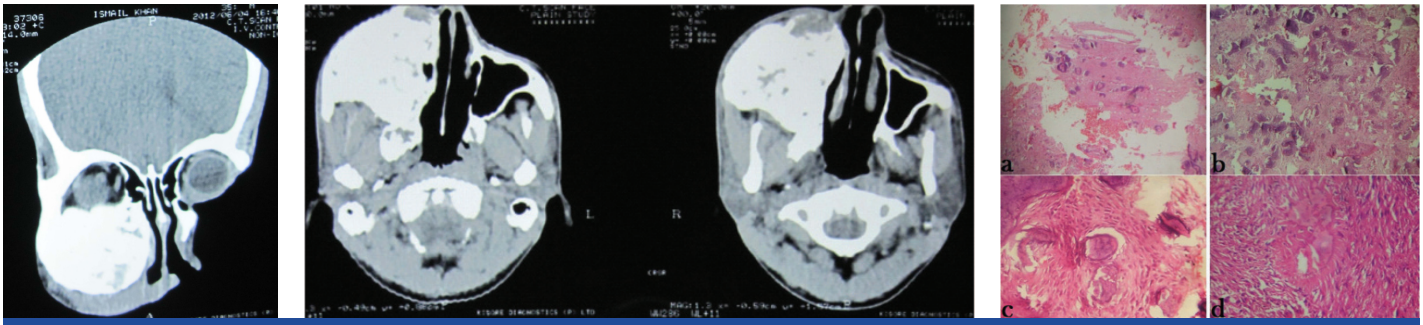
Central ossifying fibroma (COF) is a benign neoplasm designated as a fibro-osseous lesion by Eversole [1]. A bewildering variety of neoplasms, both benign and malignant, are encountered in the maxillofacial region which pose a diagnostic challenge owing to the complexity of the anatomical structures and diversity of odontogenic and non-odontogenic lesions arising in the region. A pathologic



**[Table/Fig-1]:** Frontal picture showing diffuse swelling on the right side of the face obliterating the nasolabial sulcus, extending up to the bridge of the nose and angle of the mouth **[Table/Fig-2a,b]:** Profile pictures showing facial swelling on the right side and normal facial profile on the left **[Table/Fig-3]:** Intra-oral swelling with expansion of the buccal cortex, obliterating the buccal sulcus. Presence of intra-oral pus discharging sinus (arrow) from alveolar sulcus in the periapical region of 14 which has extensive gingival recession buccally with flaring of root **[Table/Fig-4]:** Maxillary right lateral occlusal radiograph showed biortical expansion with patchy radio-opacities giving a 'cotton wool' appearance



**[Table/Fig-5a,b]:** Postero-anterior view (5a) and Water's view (5b) of skull showing a localized, well circumscribed mixed radio-opaque-radiolucent lesion in the right maxilla obscuring the antrum and displacing the orbital floor **[Table/Fig-6]:** Panoramic radiograph showing a localized, well defined, ovoid mixed predominantly radiopaque-radiolucent lesion with expansion of the antral walls **[Table/Fig-7]:** Coronal CT images of maxilla showing a well-defined, hyperdense, space occupying, and expansile lesion in the right maxilla, with bicortical expansion and displacement of the floor of the orbit and the nasal cavity, entirely filling up the right maxillary antrum



**[Table/Fig-8]:** Coronal CT scan of maxilla showing a well-defined, heterodense, space occupying, and expansile lesion in the right maxilla, with bicortical expansion, perforation of buccal cortex (arrow) and displacement of the floor of the orbit and the nasal cavity, entirely filling up the right maxillary antrum **[Table/Fig-9]:** Axial CT scan of maxillofacial region showing a well-defined, hyperdense, space occupying, and expansile lesion in the right maxilla, breaching the soft tissues buccally **[Table/Fig-10]:** Haematoxylin-Eosin stained micro section of the tumour revealed connective tissue stroma with cementum like areas at 4x magnification (10a) and 10x magnification (10b), spindle and stellate shaped cells at 40x magnification(10c) interspersed with osteoid like areas at 40x magnification (10d).

process in which the normal bone architecture is substituted by fibrous tissue containing variable amounts of mineralized material leads to the formation of Fibro-osseous lesions (FOL) [1]. FOL of the maxillofacial bones comprise of a diverse group of lesions which may be developmental, reactive or neoplastic [Table/Fig-11]. Irrespective of the type of FOL, all exhibit replacement of normal bone by fibrous connective tissue with some type of mineralized substance which may be mature or immature bone, cementum or osteo-cementum like calcifications.

In 1872 Menzel first described Cemento-ossifying fibroma but it was Montgomery in the year 1927 who coined the term OF. It was thought that Fibrous dysplasia (FD) and OF were variants of the same entity till 1948 when Sherman and Sternberg described OF in detail and since then OF has been considered as a distinct lesion [2].

The WHO first classified cementum containing lesions to be of four types: Fibrous dysplasia, ossifying fibroma, cementifying fibroma and cemento-ossifying fibroma. The next WHO classification categorized fibro-osseous lesions of the maxillofacial region into two types: osteogenic neoplasms and non-neoplastic bone lesions. In 1992, the WHO further revised the nomenclature of the separate neoplasms of ossifying fibroma and cementifying fibroma as a single entity of "cemento-ossifying fibroma". The latest 2005 classification of the WHO now classifies cemento-ossifying fibroma as a benign fibro-osseous neoplasm which is included among the non-odontogenic tumours, arising from the mesenchymal blast cells of the periodontal ligament, with a potential to form either fibrous tissue, bone, cementum, or a combination of such tissues [3]. The terms "cementifying, cemento-ossifying and ossifying fibroma" were reduced to a single term Ossifying Fibroma.

The origin of COF has been attributed to the periodontium as it has the potential to form both cementum and bone. Chronic inflammation and fibrosis of the periodontium due to periapical infection or injury to the periodontium may stimulate the formation of the lesion. But the pathogenesis of this lesion is controversial as neoplastic lesions of similar histology have been reported from other bones of the face and the long bones which lack the periodontal ligament and sometimes referred to as 'cementiform fibrous dysplasia'. It has been hypothesized by Brademann et al., that primitive mesenchymal cells in the other bones may differentiate into periodontal ligament and local trauma may be a factor in the induction of proliferation of COF [4].

All the FOL share some common clinical characteristics like slow growth and lack of any symptoms. As the lesions enlarge, they cause cortical expansion leading to facial asymmetry. If the tumour mass impinges on any nerve there may be pain or paraesthesia. Teeth

| |
|---|
| **I. Bone dysplasias** |
| a. Fibrous dysplasia<br>i. Monostotic<br>ii. Polyostotic<br>iii. Polyostotic with endocrinopathy (McCune-Albright)<br>ivOsteofibrous dysplasia<br>b. Osteitisdeformans<br>c. Pagetoid heritable bone dysplasias of childhood<br>d. Segmental odontomaxillary dysplasia |
| **II. Cemento-osseous dysplasias** |
| a. Focal cemento-osseous dysplasia<br>b. Florid cemento-osseous dysplasia |
| **III. Inflammatory/reactive processes** |
| a. Focal sclerosing osteomyelitis<br>b. Diffuse sclerosing osteomyelitis<br>c. Proliferative periostitis |
| **IV. Metabolic Disease: hyperparathyroidism** |
| **V. Neoplastic lesions (Ossifying fibromas)** |
| a. Ossifying fibroma NOS<br>b. Hyperparathyroidism jaw lesion syndrome<br>c. Juvenile ossifying fibroma<br>i. Trabecular type<br>ii. Psammomatoid type<br>c. Gigantiform cementomas |

**[Table/Fig-11]:** Classification of benign fibro-osseous lesions of the craniofacial complex

in the vicinity of the lesion retain their vitality and root resorption is uncommon. COF commonly occurs between the third and fourth decades of life and shows a strong female gender predilection of 5:1. In about 70-90% of all cases the mandible is affected [5]. We have used the term 'giant ossifying fibroma' as the tumour in our case measured more than 8cm in greatest diameter and is the largest to be reported in the maxilla of a male patient [6]. The presence of an intra-oral pus discharging sinus was suggestive of secondary bacterial infection as the tumour mass was in close vicinity of the oral mucosa.

The radiologic appearance of COF is variable, depending on the degree of mineralization of the lesion. A well-defined unilocular or multilocular lesion is seen with an internal structure which is entirely radiolucent or a mixed radiopaque-radiolucent appearance [7]. Sometimes a radiolucent line is seen in the periphery of the lesion suggestive of a capsule and is referred to as 'Rind sign' [8]. Generally teeth or roots are displaced but root resorption is uncommon. CT scan is useful for delineating the extent of the lesion. The present case exhibited entire filling up of the antrum with expansion of its walls.

Histological picture exhibits hypercellularity of fibrous stroma interspersed with calcified islands of osteoid, bone or cementum. The bony trabeculae are of variable sizes showing both lamellar and woven patterns. Peripheral osteoid and osteoblastic rimming are seen [9]. Peripheral brush borders are seen blending into the connective tissue from the spherules of cementum like material. But, there is controversy regarding the histopathologic features of OF and FD [10]. The only distinct feature of OF is presence of a fibrous capsule. As the lesion matures the calcific deposits increase but no cellular atypia or mitotic figures are seen.

As COF and FD resemble each other in their clinical, radiological as well as histopathologic features both need to be differentiated during diagnosis. [Table/Fig-12] summarizes the comparision between both entities.

Since COF is well circumscribed and well demarcated from the surrounding bone, complete surgical removal poses no problems in case of small lesions, especially in the mandible. In case of maxillary tumours as the present case filling the antrum, complete surgical removal is challenging [11]. The recurrence rate after surgical removal is about 6-28% which further increases for maxillary lesions. Despite their tendency for local invasion and recurrence, COF carries a good prognosis.

| No | Criteria | Central Ossifying Fibroma | Craniofacial Fibrous Dysplasia |
|---|---|---|---|
| 1 | Type of Lesion | Benign Neoplasm | Bone dysplasia |
| 2 | Aetiopathogenesis | Hypothesized to originate from Periodontium following infection/injury | Genetic disorder occurs due to mutation in the GNAS1 gene |
| 3 | Age | 3rd to 4th decade | 1st to 2nd decade |
| 4 | Gender | Female | No gender predilection |
| 5 | Common site | Premolar-molar region, mandible | Maxilla |
| 6 | Clinical manifestation | Painless swelling, facial asymmetry | Painless swelling, facial asymmetry |
| 7 | Radiographic appearance | Well demarcated unilocular/multilocular lesion with calcifications, Cortical expansion without perforation. | Radiolucent lytic lesions with a homogenous ground-glass appearance and ill-defined borders. |
| 8 | Histologic features | Fibrous capsule, bony trabeculae with osteoblastic rimming in hypercellular stroma with deposits of osteoid and cementum. | Irregularly shaped trabeculae of immature (woven) bone in a Chinese letter pattern in a cellular, loosely arranged fibrous stroma. |
| 9 | Treatment | Surgical enucleation | Surgical recontouring for aesthetic reasons |
| 10 | Prognosis | Good. Larger lesions may have local invasion and recurrence. | Good |

**[Table/Fig-12]:** Comparison between central ossifying fibroma and fibrous dysplasia

A comparison was done with five recently published case reports of COF as detected in a PubMed search [12-16]. It was seen that presence of a swelling and facial asymmetry was the common complaint, the age ranged from 11 to 80 y with a definite female gender predilection with a ratio of 4:1 and the mandible was the commonly affected site. Clinically there was the presence of a well circumscribed, non-tender bony hard swelling and radiographic appearance was similar revealing a well-defined expansile lesion consisting of a mixed of radiolucent-radiopaque internal structure. Though two cases had huge lesions of more than 10cm diameter [15,16], none had secondary infection with the presence of an intra-oral pus discharging sinus which makes the present case unique.

## CONCLUSION

Central ossifying fibroma is a benign fibro-osseous neoplasm which occurs as a mixed radiopaque-radiolucent lesion. Rarely such huge lesions as the present case are encountered. However, it is essential to diagnose differentiating it from other odontogenic lesions and follow up over a long term.

## REFERENCES


[1] Eversole R, Su L, ElMofty S. Benign fibro-osseous lesions of the craniofacial complex. A review. *Head Neck Pathol*. 2008;2:177–202.
[2] Worawongvasu R, Songkampol K. Fibro-osseous lesions of the jaws: an analysis of 122 cases in Thailand. *J Oral Pathol Med.* 2010;39:703–08.
[3] Barnes L, Everson J, Reichart P. World Health Organization Classification of Tumours: Pathology and Genetics of Tumours of the Head and Neck. Lyon, France, *International Agency for Research on Cancer,* 2005.
[4] Brademann G, Werner JA, Janig U, et al. Cemento-ossifying fibroma of the petromastoid region. Case report and review of the literature. *J Laryngol Otol.* 1997;111:152-55.
[5] Rajendran R. Benign and malignant tumors of the oral cavity. In: Rajendran R, Sivapathasundaram B, editors. *Shafer's textbook of oral pathology.* 6th ed. Elsevier; 2009. p. 129–30.
[6] Brannon RB, Fowler CB. Benign fibro-osseous lesions: a review of current concepts. *Adv Anat Pathol*. 2001;883:126–43.
[7] Mithra R, Baskaran P, Sathyakumar M. Imaging in the Diagnosis of Cemento-Ossifying Fibroma: A Case Series. *J Clin Imaging Sci.* 2012;2:52.
[8] Liu Y, You M,Wang H, et al. Ossifying fibromas of the jaw bone: 20 cases. *Dentomaxillo facial Radiol.* 2010;39:57–63.
[9] Naik RM, Guruprasad Y, Sujatha D, et al. Giant cemento-ossifying fibroma of the mandible. *J Nat Sc Biol Med*. 2014;5:190-94.
[10] de Andrade M, Silva-Sousa YTC, Marques MFT, et al. Ossifying Fibroma of the Jaws: A Clinicopathological Case Series Study. *Brazilian Dental Journal.* 2013;24:662-66.

[11] Gondivkar SM, Gadbail AR, Chole R, et al. Ossifying fibromas of the jaws: Report of two cases and literature review. *Oral Oncol.* 2011;47:804-09.
[12] Khan SA, Sharma NK, Raj V, Sethi T. Ossifying fibroma of maxilla in a male child: Report of a case and review of the literature. *Natl J Maxillofac Surg.* 2011;2:73-79.
[13]` Sheikhi M, Mosavat F, Jalalian F, Rashidipoor R. Central cementifying fibroma of maxilla. *Dent Res J (Isfahan).* 2013;10(1):122-25.
[14] Charavarty S, Datta D, Boro E, Dutta B. Ossifying fibroma in the sinonasal tract: a rare case report. *J Clin Diagn Res.* 2014;8(8):FD09-10.
[15] Saikrishna D, Shetty S, Ramya S. Massive ossifying fibroma of mandible. *Ann Maxillofac Surg.* 2014;4:81-84.
[16] Ram H, Mohammad S, Husain N, Malkunje LR. Huge ossifying fibroma of the maxilla. *J Maxillofac Oral Surg.* 2014;13(3):337-40.

**PARTICULARS OF CONTRIBUTORS:**
1. Reader, Department of Oral Medicine & Radiology, Institute of Dental Sciences, Bhubaneswar, Odisha, India.
2. Reader, Department of Oral Medicine & Radiology, Dasmesh Institute of Research & Dental Sciences, Talwandi Road, Faridkot, Punjab, India.
3. Reader, Department of Oral Pathology & Microbiology, Kalka Dental College, Meerut, Uttar Pradesh, India.
4. Senior Lecturer, Department of Oral Medicine & Radiology, Institute of Dental Sciences, Bhubaneswar, Odisha, India.
5. Senior Lecturer, Department of Oral Medicine & Radiology, Institute of Dental Sciences, Bhubaneswar, Odisha, India.

**NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:**
Dr. Satya Ranjan Misra,
Prasanti' Kathogola Road, Mangalabag, Cuttack-753001, Odisha, India.
E-mai : drsatyaranjanmds@gmail.com

**FINANCIAL OR OTHER COMPETING INTERESTS:** None.

Date of Submission: **Sep 10, 2014**
Date of Peer Review: **Oct 16, 2014**
Date of Acceptance: **Oct 30, 2014**
Date of Publishing: **Jan 01, 2015**