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Case Series

Radiology Section

Computed Tomographic findings in Patients with Craniofacial Fibrous Dysplasia: A Case Series

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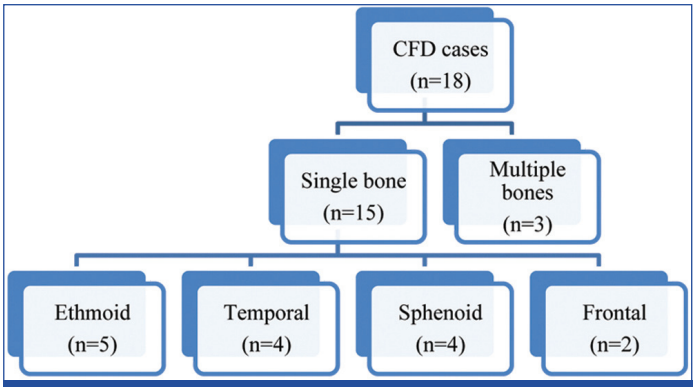
ABSTRACT

Craniofacial Fibrous Dysplasia (CFD) is a rare developmental non inheritable, non neoplastic fibro-osseous disease of the bone, which can be monostotic or polyostotic. The term CFD is used to describe FD where the lesions are confined to contiguous bones of the craniofacial skeleton. This is a case series of 18 patients with features of FD in facial and skull bones on Computed Tomography (CT). CT findings of CFD in relation to lesion location, number, appearance, and gender of the patient were documented. Out of the 18 patients, 10 were males and eight were females. The mean age was 36.4 years with a range of 13 to 77 years. Single bone involvement was seen in 15 patients, and multiple bone involvement was seen in three patients. Among the cases with single bone involvement, the ethmoid bone was involved in the majority of the cases (n=5), while the sphenoid and temporal bones were equally involved in four patients. Two cases showed lesions in the frontal bone. Ground glass attenuation was seen in the majority (n=13) of the cases. Four cases showed mixed attenuation, and sclerosis was seen in one patient. CT is the investigation of choice for craniofacial dysplasia, which can help in the diagnosis and extent of the disease. Apart from diagnosis, CT is helpful in monitoring progression and treatment planning of the disease. CT is the first investigation of choice for craniofacial dysplasia since a plain radiograph of the skull can show a complex appearance due to overlapping structures.

Keywords: Ethmoid bone, Fibro-osseous disease, Ground glass attenuation

INTRODUCTION

FD is a fibro-osseous lesion affecting the bony structures of the body. It is a non inheritable, non neoplastic developmental bone disease. Within FD, cancellous bone is replaced by abnormal fibrous and osseous tissues. FD can manifest as either monostotic (affecting one bone) or polyostotic (affecting multiple bones), with respective incidences of 70% and 30% [1]. Craniofacial involvement occurs in approximately 100% of polyostotic cases and 30% of monostotic cases [2]. FD can also be associated with syndromes such as McCune-Albright syndrome, characterised by a triad of café-au-lait skin lesions, precocious puberty, and FD, as well as MazaBraud syndrome, which presents with multiple soft tissue myxomas alongside FD [3,4].



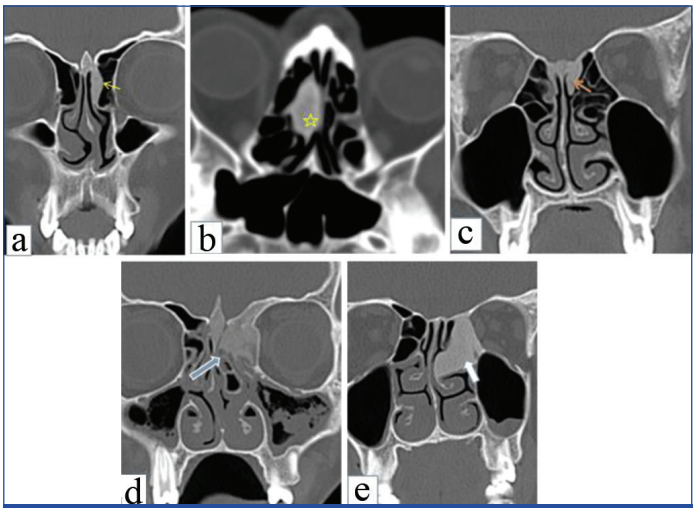
[Table/Fig-1]: Showing the location of the CFD cases.

CASE SERIES

This case series involved 18 patients who exhibited FD in the facial and skull bones on CT scans. Imaging was done using 16-slice CT equipment, specifically the SOMATOM Emotion eco, with the acquisition of axial and coronal slices. The slice thickness and table increment were 3 mm and 5 mm, respectively. CT findings of CFD were documented in relation to lesion location, number, appearance, and the gender of the patients. None of the patients had any skin lesions such as café-au-lait macules, and there were no cases of McCune-Albright syndrome. Additionally, there was no significant history of endocrinopathy, cancer, radiation therapy, facial surgery, neurosurgery, or facial or cranial trauma among the cases.

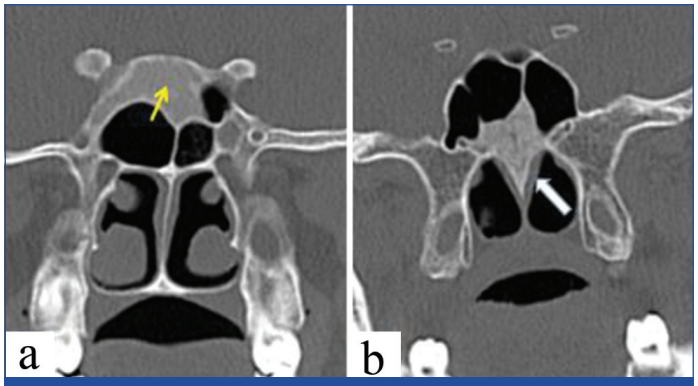
Out of the 18 patients, 10 were males and eight were females. The mean age was 36.44±17.14 years, with a range of 13 to 77 years. Headache was the most common clinical symptom in the majority of cases (n=9). Four patients experienced nasal blockage, and one patient presented with swelling and facial deformity. Incidental detection of CFD was found in four patients.

Single bone involvement was observed in 15 patients, while multiple bone involvement was seen in three patients [Table/Fig-1]. Among the cases with single bone involvement, the ethmoid bone [Table/Fig-2] was involved in the majority of cases (n=5), while

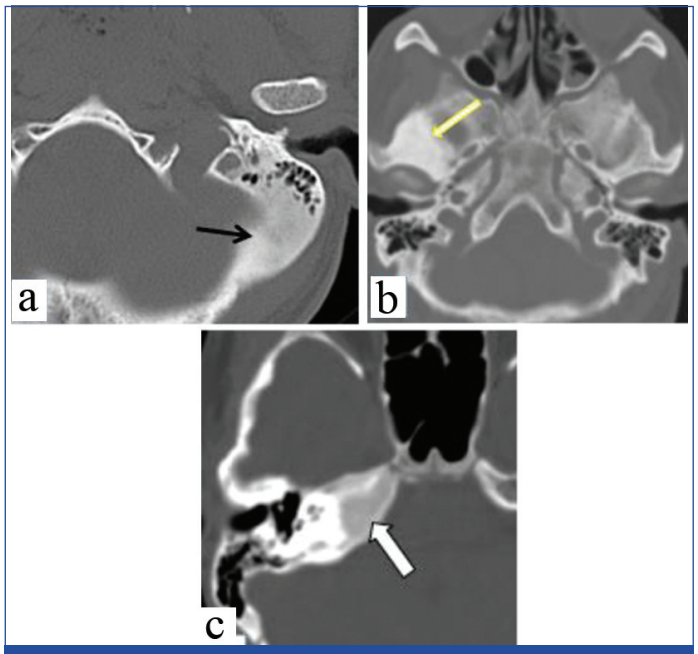


[Table/Fig-2]: (a-e): Coronal CT scan of the paranasal sinuses in bone window showing FD involving left ethmoidal trabeculae (yellow arrow in a). Axial CT scan of the paranasal sinuses showing fibrous dyplasia involvement of right ethmoidal trabeculae (yellow star in b). Coronal CT scan of the paranasal sinuses in bone window showing involvement of superior turbinates (orange arrow in c), ethmoidal trabeculae and crista galli (blue arrow in d) and left middle turbinate (white thick arrow in e) by FD.

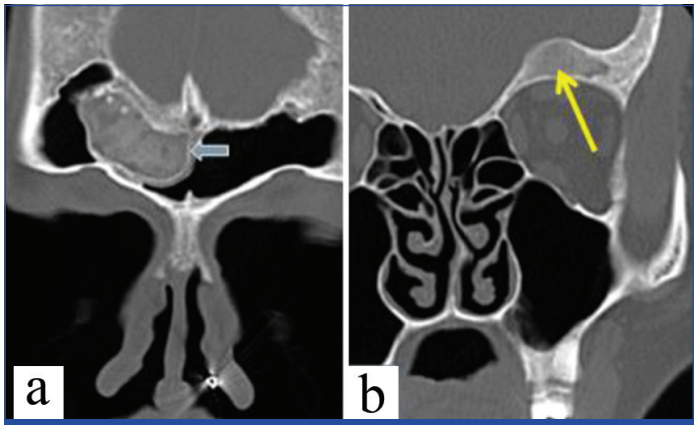
the sphenoid [Table/Fig-3] and temporal bones [Table/Fig-4] were equally involved in four patients. Two cases exhibited lesions in the frontal bone [Table/Fig-5]. CT scans without intravenous contrast revealed ground glass attenuation [Table/Fig-2,3] in the majority of cases (n=13). Four cases showed mixed attenuation [Table/Fig-6], and sclerosis [Table/Fig-4b] was observed in one patient. Biopsy was performed in two patients, confirming a fibro-osseous lesion. Histopathological diagnosis was not available for the remaining patients. There was one case where the clivus was involved and crossed the sphenoid-occipital suture to affect the petrous bone, as demonstrated in [Table/Fig-7]. None of patients underwent surgical treatment. All of them were treated conservatively. Symptomatic treatment was given.



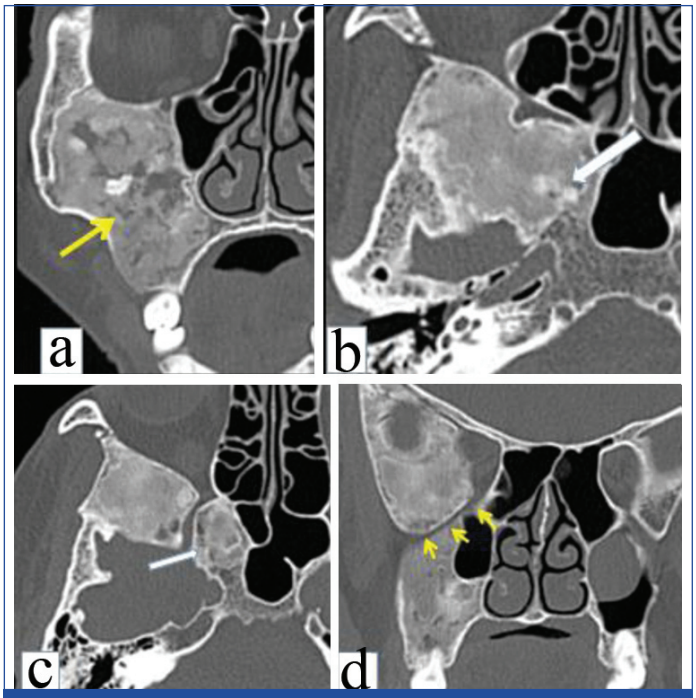
[Table/Fig-3]: Coronal CT scan images in bone window showing expansion with ground glass density of the sphenoid bones, yellow arrow in a showing involvement of roof of sphenoid bone and thick white arrow in b showing involvement of base and posterior part of vomer bone.



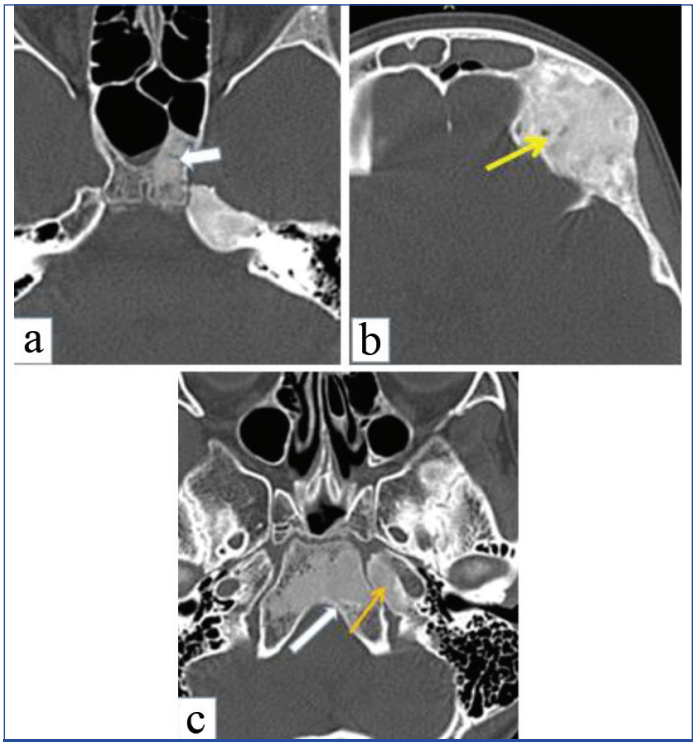
[Table/Fig-4]: Axial CT scan of the brain in bone window showing FD in mastoid segment of left temporal bone (black arrow in a), squamous part of right temporal bone (yellow arrow in b) and petrous part of right temporal bone (thick white arrow in c). Notice the diffuse sclerotic appearance of FD in b as marked by yellow arrow.



[Table/Fig-5]: Coronal CT scan of paranasal sinus in two different patients with FD showing involvement of right frontal bone in a (marked by blue arrow) and left frontal bone in b (shown by yellow arrow).



[Table/Fig-6]: Coronal and axial CT scan of the paranasal sinus of a 55-year-old lady who had presented with facial swelling and asymmetry showed fibro-osseous lesion involving maxillary bone (yellow arrow in a), greater wing of sphenoid bone (thick white arrow in b), pterygoid plate (white arrow in c) causing narrowing of inferior orbital fissure (yellow arrows in d). Note the mixed densities (ground glass, sclerosis and lucent areas) of this lesion. Biopsy of this lesion showed fibro-osseous lesion.



[Table/Fig-7]: Axial CT scan of the skull in bone window depicting the involvement of multiple bones in this patient of CFD. Thick white arrow in a showing involvement of the left side of sphenoid bone, and the yellow arrow in b shows the involvement of the left frontal bone. Figure c shows the involvement of the clivus (Thick white arrow) and the petrous part of the left temporal bone (orange arrow).

All the patients with their symptoms and CT findings are summarised in [Table/Fig-8].

DISCUSSION

FD has an incidence of 1:4,000-1:10,000 [5]. CFD primarily affects the bones of the craniofacial complex, including the mandible,

S. No.	Age (years)	Gender	Clinical details	Appearance on CT	Lesion location
1.	65	F	Headache	Mixed	Right frontal sinus
2.	18	M	Headache	GGO	Squamous part of left temporal bone
3.	40	M	Incidentally detected	GGO	Mastoid segment of left temporal bone incidentally detected
4.	42	M	Nasal block	GGO	Left anterior ethmoid sinus and crista galli
5.	36	M	Headache	GGO	Right petrous bone
6.	42	F	Incidentally detected	GGO	Right superior turbinate incidentally detected
7.	34	M	Nasal block	GGO	Post part of vomer and basisphenoid
8.	77	M	Headache	Sclerosis	Squamous part of temporal
9.	24	M	Nasal block	GGO	Left superior turbinate
10.	28	F	Headache	GGO	Roof of sphenoid sinus
11.	49	F	Headache	GGO	Roof of left frontal sinus
12.	19	F	Headache	GGO	Right ethmoid sinus and middle turbiante
13.	26	M	Incidentally detected	GGO	Left ethmoid sinus and middle turbinate
14.	19	F	Headache	Mixed	Bilateral frontal bones, sphenoid bones and crista galli
15.	13	M	Incidentally detected	GGO	Left frontal bone, roof of left orbit, clivus, basisphenoid on left side and left petrous bone
16.	27	F	Nasal block	GGO	Left ethmoidal trabeculae
17.	42	M	Headache	Mixed	Sphenoid sinus
18.	55	F	Swelling	Mixed	Right maxillary bone, greater wing of right sphenoid bone and orbit

[Table/Fig-8]: Summary of all 18 cases.
M: Male; F: Female; GGO: Ground glass attenuation

maxilla, cranial base, and vault [6]. FD is most commonly observed in children and young adults, although it can sometimes go unnoticed until middle age. A slight female preference has been observed [5]. The maxilla (12%) and mandible (12%) are the bones most commonly affected, while involvement of the ethmoid, sphenoid, frontal, and temporal bones is less frequent. However, in this series, the ethmoid bone was the most frequently affected, being involved in five patients. This aligns with a study conducted by Lustig LR et al., which found that the ethmoid bone was the most commonly affected (71%) [7]. CFD primarily involves the bones of the face and skull, with the parietal and occipital bones being less affected [8]. Lesions of the clivus are extremely rare [9]. FD can cross sutures without any interruption [10].

The clinical presentation of CFD varies depending on the affected bone and its proximity to nearby structures such as foramina. Small lesions of the skull vault may be asymptomatic and incidentally detected on CT scans. Large expansile lesions of the skull vault can cause swelling [8]. The literature describes three different radiographic patterns of CFD, including a mixed lucent and sclerotic pattern (pagetoid type) observed in 56% of cases, a homogeneous sclerotic pattern in 21% of cases, and a predominantly cystic morphology with round or oval lucency surrounded by a dense sclerotic boundary in 21% of cases [11]. In this series, authors encountered an elderly patient showing a mixed pattern of attenuation on CT scans. This was consistent with a study conducted by Kushchayeva YS et al., which states that craniofacial lesions in older individuals tend to become less homogeneous on CT scans, developing discrete radiolucent, cystic-appearing areas [12].

An accurate diagnosis of craniofacial dysplasia on a radiograph cannot be made due to the complex and overlapping structures of the facial skeleton. Hence, CT is the preferred choice for diagnosing craniofacial FD [10,13,14]. CT features of FD include bone expansion with variable Hounsfield units ranging from 34 to 513 HU, depending on the proportions of fibrous tissue and bone content [6]. CT provides precise measurements of the extent, dimensions, and radiodensity of FD, allowing for evaluation of long-term changes and progression [15]. Both CT and Magnetic Resonance Imaging (MRI) are effective in assessing the impact of CFD on neurovascular structures, orbit, optic canals, and adjacent paranasal sinuses. CT is useful for evaluating both bony and soft tissue details, while MRI is more sensitive to pathological changes but can be challenging to interpret due to FD's low signal intensity [16]. Therefore, CT is the preferred modality for investigating CFD [16].

Similar to the study by Yanez N et al., present study did not found cases describing the presence of craniofacial lesions inconjunction with lesions in other body parts, whether monostotic or polyostotic [17].

The management of facial lesions depends on the patient's stage of development and skeletal maturity. Surgical excision is the only technique that achieves complete resolution of FD. For stable lesions that cease to grow after puberty, a wait-and-watch approach may be appropriate.

CONCLUSION(S)

CT is the first investigation of choice for craniofacial dysplasia, as a plain radiograph of the skull can exhibit a complex appearance due to overlapping structures. CT is a valuable tool in CFD cases for demonstrating the extent of the disease, evaluating neural and vascular foramina, and monitoring patient progress.

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