

## GROUND-GLASS PATTERN FIBROUS DYSPLASIA OF FRONTAL SINUS

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**The case of a 5-year-old boy with ground-glass pattern craniofacial fibrous dysplasia (FD) presenting with progressive swelling in the right frontal region is reported. The imaging findings with computed tomography and magnetic resonance imaging (MRI) findings are presented. The differential diagnosis with inspissated mucocele is discussed as well.**

**Fibrous tissue could be hypo dense on CT. Post-contrast enhancement of the lesion on MRI is the key for diagnosis.**

**Key-word: Paranasal sinuses.**

Fibrous dysplasia (FD) is a non-inherited benign developmental skeletal disorder, in which abnormal differentiation of osteoblasts leads to replacement of normal marrow and cancellous bone by immature bone and fibrous stroma. In case of monostotic, fibrous dysplasia of craniofacial bones the frontal and sphenoid bones are most commonly involved. We report a new case of ground-glass pattern fibrous dysplasia of frontal sinus in a 5-year old boy, mimicking inspissated mucocele on CT.

### Case report

A 5-year-old boy presented with a 2 year history of progressive swelling in the right frontal region. His medical history was unremarkable. Physical examination revealed protrusion in the right frontal region and slight proptosis, however with no effect at the visual ability. Computed tomography (CT) demonstrated a large lesion expanding the frontal sinus, involving the nasal cavity and ipsilateral ethmoid sinuses as well. The lesion had well defined margins, and it was hypodense in the range of water. A calcified area was depicted centrally (Fig. 1). The right ocular bulb was displaced anteroinferiorly. The differential diagnosis of the lesion on CT was between an expanded airless sinus cavity filled with inspissated mucocele and a benign fibrous lesions of frontoethmoid area.

On MRI the lesion demonstrated low-signal intensity on both T1- and T2- weighted spin-echo sequences, and enhanced markedly after intravenous injection of Gd-DTPA. The calcified area showed an even lower

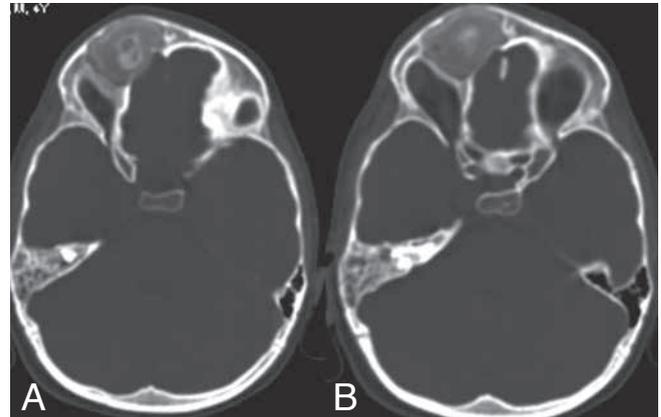


Fig. 1. — Computed tomography with bone window images (A, B) shows a hypo dense lesion with well defined margins expanding the right frontal sinus. A calcified area is seen centrally.

signal intensity on both T1- and T2-weighted SE sequences (Fig. 2). The enhancement of tissue filling the frontal sinus with the additional presence of central sclerotic lesion favoured the diagnosis of benign fibrous lesions of frontoethmoid area.

The patient was referred to surgery with a diagnosis benign fibrous lesions of frontoethmoid area, and cranioplasty was performed for excision. Specimens from lesion were sent for histological examination. The histopathology was consistent with craniofacial fibrous dysplasia.

### Discussion

Fibrous dysplasia (FD) is a non inherited benign developmental skeletal disorder of unknown origin in which abnormal differentiation of

osteoblasts leads to replacement of normal marrow and cancellous bone by immature bone and fibrous stroma. Even though it may be an incidental finding, it may be also complicated by pathologic fracture, and rarely by malignant or sarcomatous degeneration in 0.5% of cases. The commoner monostotic form of FD, characterized by single-bone involvement primarily in the extremities and ribs, tends to become quiescent with cessation of growth. The polyostotic form affects multiple bones usually in a unilateral distribution and may continue to progress beyond puberty. When polyostotic FD is associated with precocious puberty and skin pigmentations, it is termed the McCune-Albright syndrome (1). When monostotic fibrous dysplasia occurs adjacent to craniofacial bones, the result is a very rare bone disease called craniofacial fibrous dysplasia (CFD). It has been reported that FD of the paranasal air sinuses usually develops as a result of extension of the disorder into the sinuses from adjacent facial osseous tissue.

The most common clinical sign of craniofacial FD is swelling. Other reported presenting signs and

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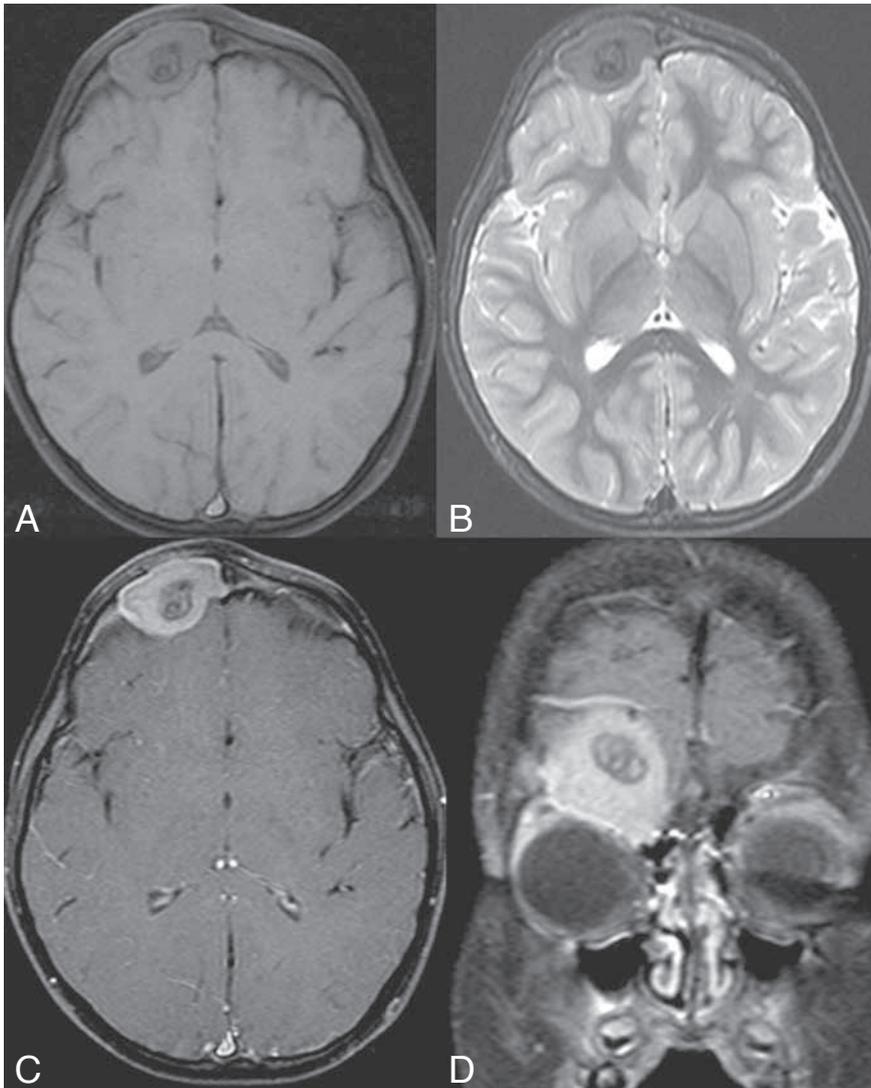


Fig. 2. — MR images: axial fat-sat T1-weighted image (A), axial STIR image (B), axial (C) and coronal (D) a post-gadolinium T1-weighted with fat saturation. The lesion demonstrated low-signal intensity on both T1- and STIR sequences, and enhanced markedly after intravenous injection of Gd-DTPA.

symptoms such as visual loss or dizziness depend on the area of involvement (2, 3, 4).

Radiologic findings in computer tomography can demonstrate the changes in the bones and are classified in three types: the ground-glass pattern (56%), the homogeneously dense pattern (23%), and the cystic variety (3, 5-7). Density levels on CT range, depending on the fibrous tissue and maturity of bone content. The sclerotic and the ground glass appearances are easily recognized as FD by the thickening of the bone. FD with a cystic appearance usually develops in the frontoethmoidal region and sometimes the cysts within the lesion cannot be differentiated from the presence of mucocele (8).

On MRI the affected bones usually have a low- to intermediate-signal intensity on both T1- and T2-weighted sequences, probably owing to the replacement of the marrow cavity by fibrous tissue. However, the lesions may have quite variable signal intensity on T2-weighted images, where they may appear hyperintense depending on the activity of the lesions. On postgadolinium MR images, affected bones show variable enhancement, reflecting vascularity of the fibrocellular tissue. MRI offers greater specificity in neurovascular and ocular involvement and in detection of other soft tissue lesions (9).

In our case, CT-scan revealed expansion of the right frontal sinus, ballooning of the affected area with a

“ground-glass” appearance. The density of the lesion was 40 HU that was less than the usually reported density of 300-600 HU of craniofacial FD. A central sclerotic area was seen as well. The lesion extended toward the orbital roof and ethmoid sinuses, had well-defined lobulated margins and it was protruding in the anterior cranial fossa compressing the frontal lobe.

On MRI the expanded frontal sinus had intermediate signal intensity on T1-weighted images and low signal intensity on T2-weighted images. The central sclerotic area showed low signal intensity on both sequences. After the administration of contrast medium intense contrast enhancement of the tissue filling the frontal sinus was depicted. Intermediated enhancement of central sclerotic area was seen as well.

The differential diagnosis of the lesion was between an expanded airless sinus cavity filled with inspissated mucocele and a benign fibroosseous lesions of frontoethmoid area.

Sinonasal secretions may have a spectrum of MR signal intensity, ranging from hyperintense to signal void with all pulse sequences. These findings must be kept in mind when interpreting images of patients with suspected chronic sinusitis. In such cases, accurate differentiation may require contrast-enhanced MR images, where most tumors show solid enhancement, unlike mucoceles, which exhibit only mural enhancement, if at all (10). In our case the enhancement of tissue filling the frontal sinus with the additional presence of central sclerotic lesion favoured the latest diagnosis.

Association of a craniofacial FD with a mucocele is a very rare occurrence that has been reported. This complication most probably results from the involvement and subsequent occlusion of the recesses of the sinuses by the dysplastic process, as it has been reported in other studies (6).

The management of fibrous dysplasia is not surgical unless it causes unacceptable or progressive deformity, cranial nerve compromise, severe headache, or development of a malignancy.

In our case despite normal vision of the patient, surgical management was chosen because of protrusion in the right frontal region and slight proptosis. Under general anesthesia, the patient underwent resection of the lesion. He exhibited marked cosmetic improvement and experienced no complications of surgery. To date, the patient has made favorable

progress and no sequela related to the surgery has occurred.

### Conclusion

The presurgical diagnosis of craniofacial fibrous dysplasia depends mainly on CT and MR scan images. Our report emphasises the radiologic appearance of frontal sinus fibrous dysplasia and the differential diagnosis with inspissated mucocele. Despite the relatively low density of fibrous tissue on CT, post-contrast enhancement on MRI was the key of the differential diagnosis. The MRI is highly sensitive in detecting enhancement of solid lesions, in mapping the relationship of a tumor with its adjacent structures and in providing sufficient correlation with histological findings.

The radiologist's goal is to be able to recognise this rare entity in order to guide treatment options.

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