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**Case Report**

# Fibrous Dysplasia of the Sphenoid and Ethmoid Bones – A Rare Benign Entity with a Destructive Potential

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## Abstract

***Background*:** Fibrous dysplasia (FD) of bone is a rare sporadic benign congenital condition in which normal bone is replaced by fibro-osseous tissue with immature osteogenesis. Sarcomatous transformation is exceptional. The most common presenting features in craniofacial forms include atypical facial pain and headaches. ***Case Presentations***: we report here 2 cases, that of a 36 year old male and of a 43 female, who both presented with a similar history of headaches. The male patient refused surgery so removal of the diseased bone was not an option. However, as the limited growth of this lesion have been established, only further, albeit close, observation was planned. The female patient underwent a successful removal of her tumour through an endoscopic approach. At the 1.5 year follow-up, she remains disease free. ***Conclusion***: craniofacial fibrous dysplasia can present itself in myriad ways, therefore, the diagnosis should always be considered in case of headache, neuralgia or sensory disorders. Modern imaging modalities and histopathologic analysis have made the diagnosis reasonably straightforward. A medico-surgical approach is useful for these patients. Surgery —when indicated— is tailored to the specific clinical presentation. The goal is to be as minimally invasive as possible while still achieving the desired outcome, prioritizing the preservation of existing function.

**Keywords:** fibrous dysplasia, sphenoid bone, ethmoid sinus, endoscopic surgery.



## INTRODUCTION

**CASE 1:**

We report the case of a non-smoker 36 year old male who was referred to the Head and Neck unit at the University College Hassan II Hospital with an 8 months history of headaches projecting bilaterally over the sphenoid region of the skull. The patient had no history of endocrinopathy, cancer, radiation therapy or surgery, be it facial or otherwise. He also had no history of injury or visual changes and no history of epistaxis. The primary physical examination was unremarkable and showed no facial or cranial deformity and or asymmetry, sensorineural disorder, functional disorder, infectious complications, signs of associated endocrinopathy or café-au-lait macules.

**Paraclinical Sensorineural Assessment:**

- Visual acuity: 10/10
- Goldmann visual field test: within normal limits.
- Audiometry: showed no hearing loss.
- Nasal endoscopy: within normal limits.

**Biological and Pathology Data:**

- Prolactin / IGF-1 / PAL / Vit-D: all within normal limits.

**Baseline Imaging:**

**Contrast Sinus Ct:**

**(Figure 1)** demonstrated an ossified mass confined to the left sphenoid sinus. There was no enhancement on IV contrast. Provisional diagnosis of fibrous dysplasia was made on CT findings.

**Contrast MRI:** ossified mass confined to the left sphenoid sinus.

The patient refused surgery so removal of the diseased bone was not an option. It was decided to keep the patient under close observation.

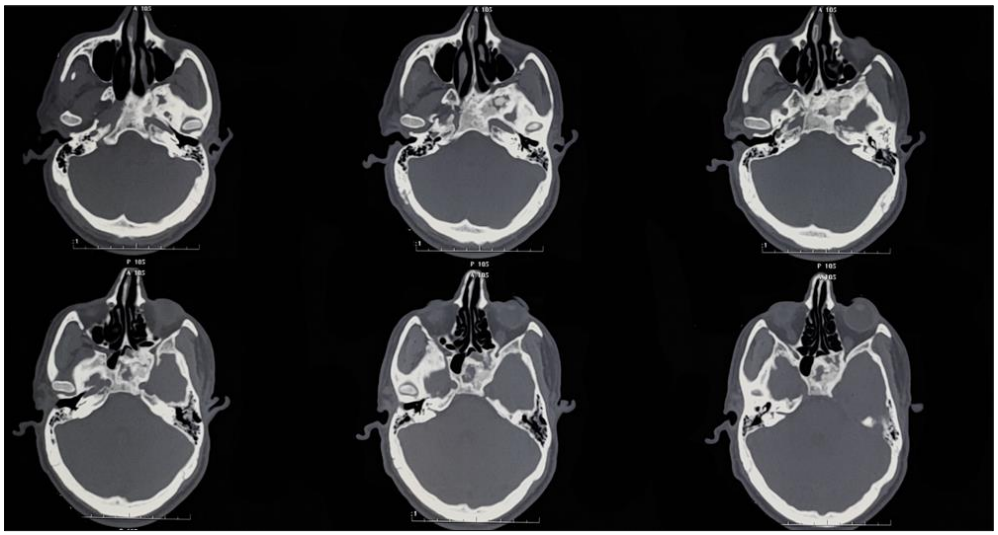
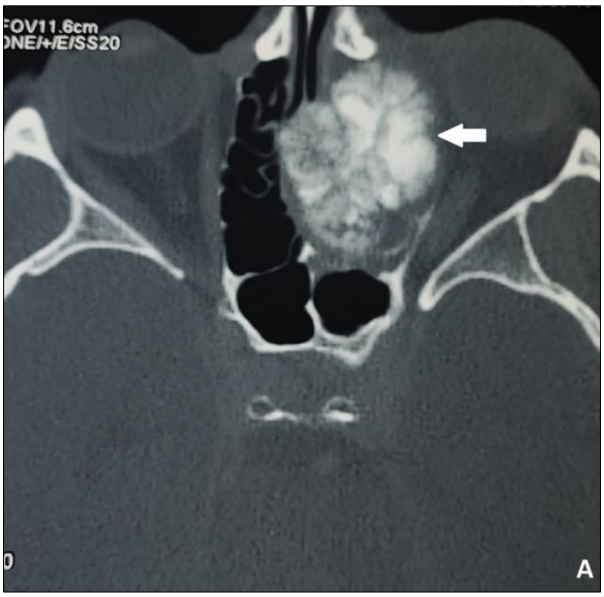
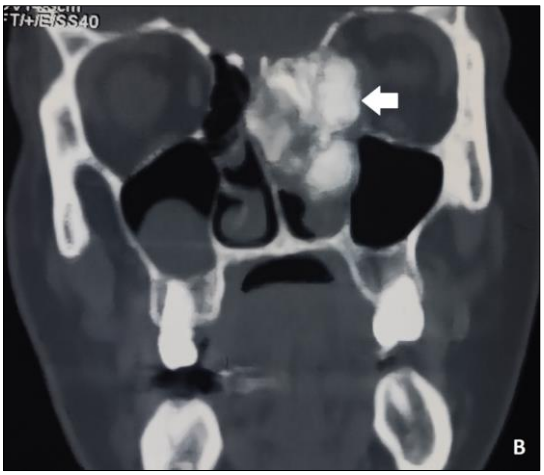


**Figure 1: Preoperative contrast sinus CT axial view of the skull base, demonstrating an ossified mass confined to the left sphenoid sinus. There was no enhancement on IV contrast.**

**CASE 2:**

A non-smoker 43 year old female presented to the Head and Neck unit at the University College Hassan II Hospital with a 12 months history of headaches projecting over the left parietal region of the skull associated with a few episodes of epistaxis. She had no history of injury or visual changes. She had no significant medical or family history. Physical examination of the patient revealed a mild pallor. Local examination showed a mild left exophthalmia. The rest of the general examination was unremarkable. Flexible naso-laryngoscopy revealed a bony mass protruding over the left nasal mid-turbinate, pushing it medially. A CT (computed tomography) scan revealed the presence of an ossified mass not confined to the left ethmoid sinus measuring approximately 36 x 30 x 45mm. That mass expanded laterally to the orbit and maxillary sinus and medially to the nasal septum. There was no enhancement on IV contrast. Provisional diagnosis of a malignant tumour was made on CT findings. Multiplanar MRI confirmed these findings (figure 2). No regional lymph nodes were otherwise detected. Paraclinical assessment showed nothing out of the ordinary. An endoscopic biopsy of the bony tumour was performed and histopathological examination was in favour of an osteoid osteoma. The patient underwent surgical resection through an endoscopic approach. The mass was carefully dissected from the adjacent structures (figure 3). The postoperative follow-up was simple allowing the patient to be discharged within 48 hours. The patient had an uneventful recovery and remained disease-free at 1, 3 and 6 months follow-ups. The symptoms had completely subsided. Histopathological analysis confirmed the benign nature of the lesion and the diagnostic of fibrous dysplasia.

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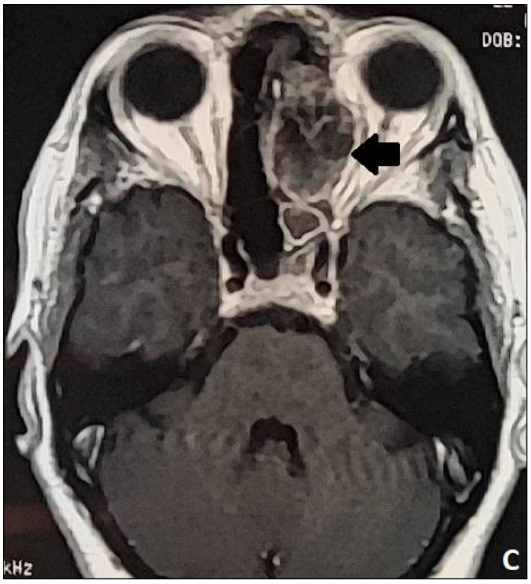
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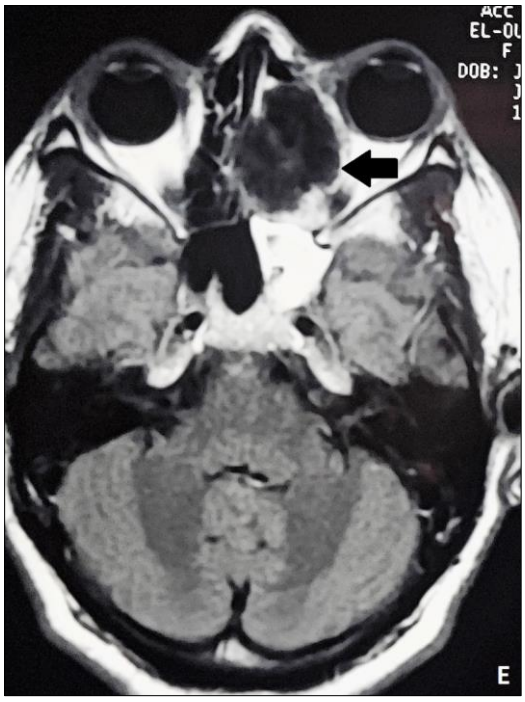
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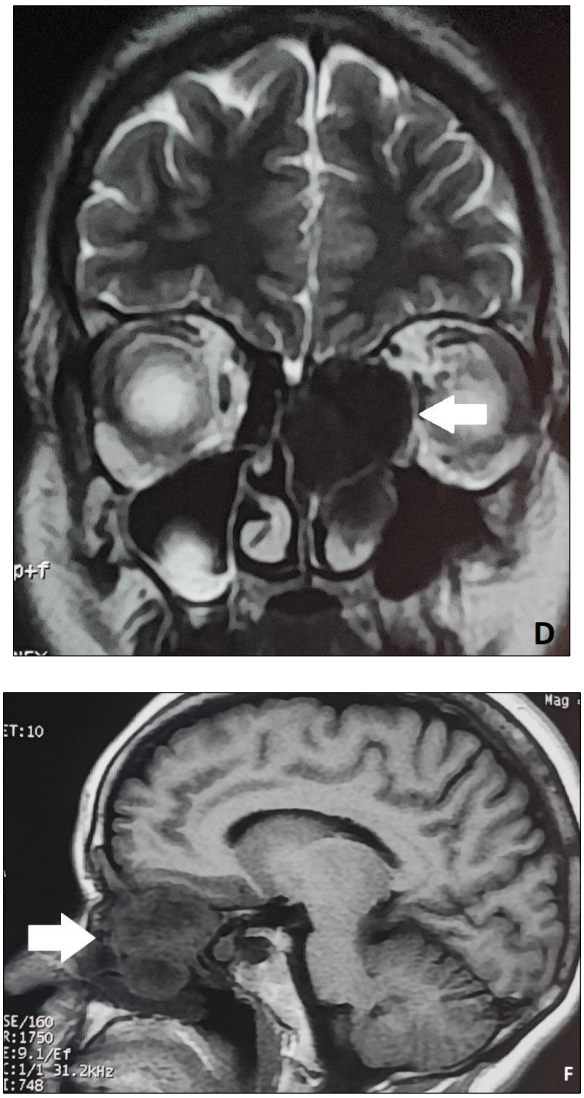


**Figures 2: Preoperative contrast sinus CT axial (2A) and coronal (2B) view of the skull base (upper row) demonstrating a heterogeneous, ossified mass of the left ethmoid sinus, expanding laterally to the orbit and maxillary sinus and medially to the nasal septum. There was no enhancement on IV contrast**

Magnetic resonance imaging axial (2C) & (2E), coronal (2D) and sagittal (2F) views of the sinuses, confirming the above-mentioned findings.

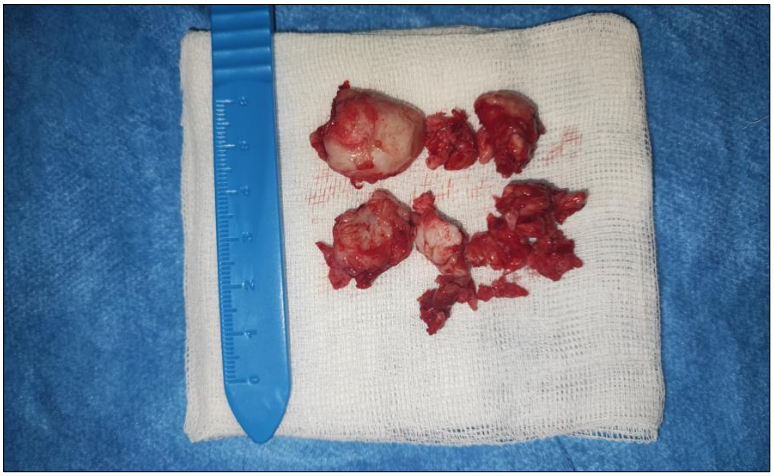

**Figure 3: surgical specimen resected through an endoscopic approach (curettage)**

## DISCUSSION

Paranasal sinus masses can be divided into benign and malignant tumors of epithelial or fibro-osseous origin. The benign fibro-osseous lesions can be further subdivided into fibrous dysplasia and ossifying fibroma.

Fibrous dysplasia of bone is a rare sporadic benign congenital condition that affects both sexes equally. ―prevalence is reported to be less than 1/2000 [1], but this number is probably underestimated due to the existence of asymptomatic forms. It can occur in one (monostotic) or multiple (polyostotic) bones with a more frequent unilateral distribution. There is no transformation from monostotic to polyostotic form, and malignant transformation (usually confined to polyostotic cases) remains a rare occurrence.

The underlying molecular abnormality concerns somatic mutation of the GNAS gene coding for the subunit of protein G, resulting in osteoblast differentiation deficit, fibrous medullary proliferation and osteoclast hyperactivity partly due to overexpression of IL-6 within mutated cells. Fibrous dysplasia typically appears in childhood and often becomes inactive by early adulthood. While the condition itself has been around for a long time, the first accurate pathological description of the disease is credited to Von Recklinghausen in 1891.

Since this same mutation can also affect other cell types, fibrous dysplasia has also been reported in association with other diseases [3], such as McCune Albright syndrome (cutaneous lesions consisting of café-au-lait macules and endocrine disorders (early puberty, hyperthyroidism, acromegaly, Cushing's syndrome) and Mazabraud's syndrome (myxomas). Craniofacial involvement is found in 50% of polyostotic and 27% of monostotic forms, mainly involving ―in order of frequency― the ethmoid (71%), sphenoid (43%), frontal bone (33%) or maxilla (29%) [2]. Other cranial bones can be affected as well, such as the temporal, parietal and occipital bones. In the vast majority of cases, sinus involvement is by direct extension from adjacent diseased bone as fibrous dysplasia prefers membranous bone.

In initial work-up, CT coupled to MRI with contrast enhancement is required for differential diagnoses and analysis of local extension and neural compression. Four radiologic differential diagnoses are to be mainly considered: skull-base en-plaque meningioma, cranial osteoma, ossifying maxillary fibroma, and Paget's disease. MRI centered on the sella turcica screens for pituitary adenoma, combined with systematic prolactin and IGF-1 assays (even in the absence of clinical evidence for endocrine involvement). Bone scintigraphy might be performed in case of atypical lesion or suspicion of malignancy.

Fibrous dysplasia is a benign tumor and therefore when asymptomatic and not cosmetically disfiguring, it can usually be monitored conservatively. When symptomatic, it's usually revealed by headache, neuralgia, sensorineural or functional disorder, infectious complications or severe esthetic blemish. When the diagnosis is unclear based on physical examination and imaging tests, a bone biopsy can prove to be helpful.

Spontaneous resolution of FD does not occur. Medical treatment is based on anti-resorption drugs (pamidronate) known for inhibiting osteoclast activity. Associating vitamin D3/calcium supplementation to bisphosphonate is recommended. Case reports suggest that tocilizumab and denosumab (monoclonal antibodies targeting the IL-6 receptor and RANKL respectively) may be beneficial treatment alternatives for patients with fibrous dysplasia not responding to bisphosphonate therapy.

Surgical intervention is indicated for symptomatic lesions causing bone pain, cosmetic deformity or local pressure on vital structures. Loss of visual acuity is the most frequent ocular complication. Other ophthalmologic symptoms include exophthalmia, epiphora, ptosis or diplopia.

Removing Fibrous dysplasia of the sphenoid and ethmoid sinuses runs the risk of injury to the skull base, carotid arteries and optic nerves and chiasm. Multiple approaches have been described, including endonasal, transfrontal and subcranial. An intranasal endoscopic approach to diagnosis allows planning of the appropriate procedure, with the comfort of an established diagnosis [4-5]. It allows a planned intracranial/extracranial approach in a surgical field that has not been encumbered by previous incisions.

Malignant transformation is rare, occurring in less than 4% of cases, depending on the series, and in either form (polyostotic or monostotic). Osteosarcoma is the most frequent malignancy, although fibrosarcoma and chondrosarcoma have also been reported. And even when a subtotal resection is performed, patients need to be controlled for years with serial scans and close follow ups to catch any recurrences.

## CONCLUSION

In case of headache, neuralgia, sensory deficit, functional disorder and/or infectious complications, craniofacial fibrous dysplasia should always be considered and facial and cranial CT and MRI should be performed. In case of pain, medical treatment is based on bisphosphonates. Surgical resection is indicated only when the disease is symptomatic in case of sensorineural disorder, and/or when for aesthetic purposes. Surveillance should be prolonged, with a sarcomatous degeneration risk, eventhough extremely rare, always to be taken into account.

**Disclosure of Interest**: the authors declare that they have no competing interest

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