

Joman Q. Laxamana, MD
Rene Louie C. Gutierrez, MD
Roberto C. Claridad, MD

Department of Otorhinolaryngology
Head and Neck Surgery
East Avenue Medical Center

Giant Ossifying Fibroma of the Frontoethmoid Sinus: A Silent Peril

ABSTRACT

Objectives: To present a rare case of a large ossifying fibroma of the frontoethmoid sinus and describe our experience with the clinical presentation, diagnosis, management dilemmas, surgical approach and outcome of our patient.

Methods:

Design: Case Report

Setting: Tertiary Government General Hospital

Patient: One

Results: A 29-year-old housewife consulted with a large left frontoethmoidal mass of 20 years duration causing significant facial deformity, left eye proptosis, headache and psychosocial distress. Initial CT scans and MRI revealed a well-encapsulated mass occupying the frontoethmoid sinus, left orbit and anterior cranial fossa and subsequent surgical management involved three important aspects: 1) Wide extirpation of the tumor; 2) Preservation of the brain, left orbital contents and function; and 3) Reconstruction of the facial defect using calvarial bone graft, abdominal fat and temporalis muscle flaps.

Conclusion: A large ossifying fibroma of the frontoethmoidal sinus threatens the integrity of the vital structures it compresses and poses compelling diagnostic and surgical challenges. Adequate imaging, multidisciplinary planning and surgical expertise are needed to ensure a successful outcome.

Keywords: *Ossifying Fibroma, frontoethmoid sinus, mucocele, orbital preservation, calvarial bone graft, abdominal fat graft*

Ossifying fibroma is a rare, benign, slow-growing neoplasm commonly found in the maxillary-mandibular area. It is infrequently found in the paranasal sinuses much less in the frontoethmoidal area. Delayed surgical management may allow these lesions grow to massive proportions and cause a variety of complications demanding specialized multidisciplinary treatment. We present one such case.

CASE REPORT

A 29-year-old single mother of three from Camarines Norte, Philippines consulted for a large left orbital mass that began 20 years ago with progressive proptosis of her left eye associated with tearing and redness with no medical consult or medications. About 16 years ago, she started to have severe episodic left sided headache and consulted a private physician. A CT scan revealed a left frontoethmoid sinus tumor with intracranial and retro-orbital extension. She underwent partial transcranial excision of the left retro-orbital mass relieving left eye proptosis and headache. Histopathology revealed Cementifying Fibroma. She was then advised complete excision of the frontal sinus tumor but was unable to comply because of personal and financial reasons. She was discharged improved after three weeks.

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Correspondence: Dr. Joman Q. Laxamana
Department of Otorhinolaryngology
Head & Neck Surgery
6th Floor, East Avenue Medical Center
East Avenue, Diliman, Quezon City 1100
Philippines
Phone: (632) 928 0611 local 324
Fax: (632) 435 6988
Email: jqlaxamana@gmail.com
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Meanwhile, the left orbital mass progressively enlarged with anteroinferior displacement of the left eyeball causing gross physical deformity and significant social and emotional distress. Remarkably, she claimed to have no blurring of vision or diplopia.

Four months before admission with a still enlarging left orbital mass and occasional mild headache she consulted at our institution and was subsequently admitted.

CT scan and MRI results revealed a 6.2 x 7.0 x 9.6cm predominantly cystic irregularly shaped mass with multiple enhancing internal septations and solid components. The mass compressed the skull base and left frontal lobe superiorly, the left maxillary sinus inferiorly, the left superior nasal turbinate medially, temporal bone laterally and the frontal and temporal lobe posteriorly. (Figure 1)

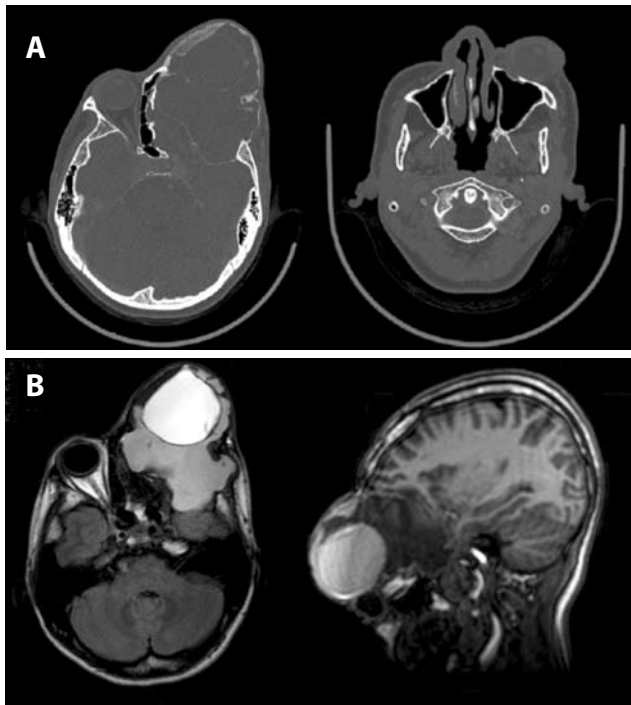


Figure 1. A. CT scan showing the frontoethmoid mass and inferoanteriorly displaced left orbit B. MRI showing the hyperintense ossifying fibroma with a surrounding expansile mucocele

Physical examination showed a 7.0 x 6.5 cm hard, fixed, non-tender left orbital mass with associated proptosis and inferior displacement of the left eye. (Figure 2) Both eyes were equally reactive to light with a visual acuity of 20/20 for the right eye and 20/125 for the left. There was moderate upward gaze deficit in the left eye.

Preoperative evaluation with 3-D rendering using Surgicase® 5.0 (Materialise, Belgium) showed a substantial defect that could cause orbital compromise, brain herniation and difficult reconstruction. (Figure 3) A virtual excision allowed accurate planning of the surgical approach minimizing intraoperative decision making.

In the operating room, a coronal incision and midfacial degloving exposed the mass from which 10cc of straw-colored fluid was aspirated. (Figure 4) The bony capsule of the mass was meticulously extirpated from



Figure 2. Preoperative Photo showing a large left frontoethmoid mass causing inferoanterior displacement of the left orbit

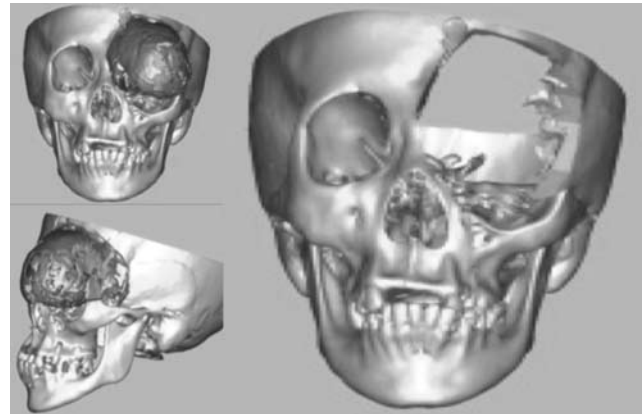


Figure 3. Preoperative planning using 3D rendering and virtual excision of the mass using Surgicase® 5.0 (Materialise, Belgium)

all its attachments preserving the frontal lobe and left eyeball yielding a 6 x 7 x 10 cm round, well delineated cystic mass with a bony capsule. (Figure 5) The orbital floor was intact but inferiorly displaced. The left optic nerve and extraocular muscles were elongated but preserved. Posteriorly, the anterior and posterior frontal tables had been eroded exposing a 4 x 3 cm portion of the frontal lobe with intact dura, which was assessed to have no need for mesh support. (Figure 6)

An abdominal fat graft was laid into the orbital defect, followed by an ipsilateral temporalis muscle sling supporting the frontal lobe. (Figure 7) A right parietal split-thickness calvarial bone graft was contoured and fixed into the left superior orbital rim. The donor site was also covered with a right temporalis muscle sling. (Figure 8) The procedure lasted six hours with a total estimated blood loss of 500cc.

Postoperative neurosurgical monitoring showed no neurologic deficits. Ophthalmologic evaluation revealed decreased proptosis (Figure 9) with the same preoperative extraocular movement on the left eye and acceptable visual acuity. The patient recovered uneventfully without any dizziness, headache or diplopia and was subsequently discharged. Final histopathological examination of the mass revealed ossifying fibroma. She was well at one-month follow-up.

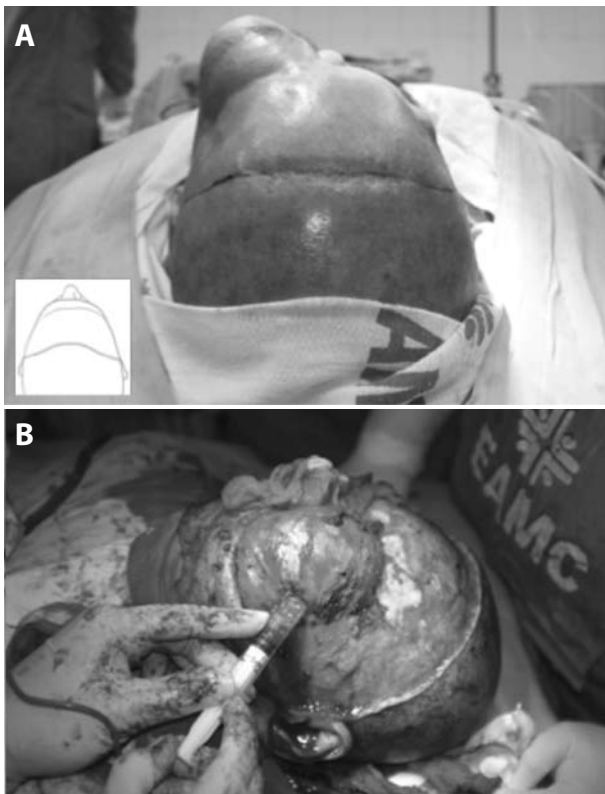


Figure 4. A. Coronal incision markings B. Aspiration of cystic contents



Figure 5. Extirpation of the bony capsule grasped using a Babcock clamp.



Figure 6. Posterior frontal table defect. The intact dura is seen shining at the posterior wall of the mass.

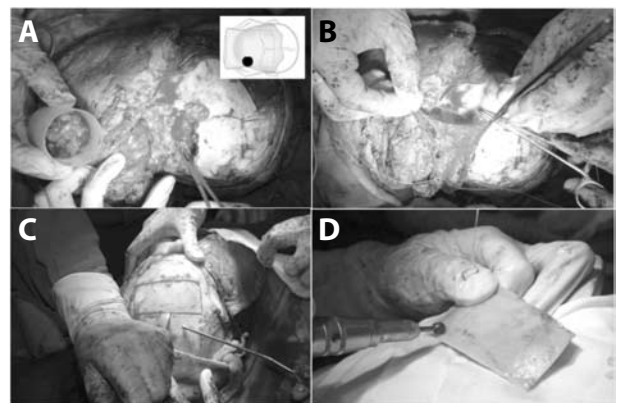


Figure 7. A. Overlaying of abdominal fat B. Temporalis muscle sling supporting the orbit C. Harvesting of calvarial bone graft D. Contouring of the graft

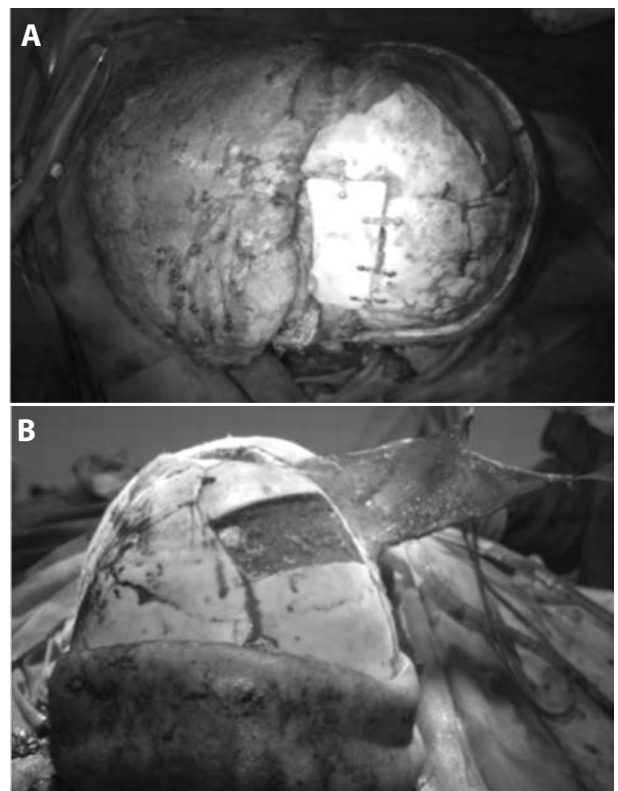
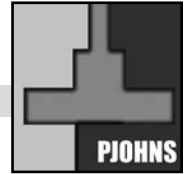


Figure 8. A. Fixation of the calvarial bone graft using titanium plates and screws B. Temporalis muscle sling over the donor site



Figure 9. Post-operative photo showing decreased proptosis



DISCUSSION

Ossifying fibroma is a benign maxillofacial fibro-osseous lesion commonly located in the mandibular molar area and rarely found in the frontoethmoid area.¹ Usually, it is neither aggressive nor excessively destructive but has a propensity for osseous cortical expansion approximately equal in all directions. It is more frequent in women than in men (4:1) in the third and fourth decade of life. Clinically, these tumors manifest as painless, well delineated, slow-growing masses that may become large and destructive over time. The prognosis is excellent after complete excision and malignant transformation has not yet been documented.² Recurrence rates are as high as 30 to 58%.³

A Google Scholar and PubMed search of MEDLINE using the keywords "ossifying fibroma," "frontoethmoid," and "frontal-ethmoid" suggests that this is the third reported case of an ossifying fibroma of the frontoethmoid sinus in the English literature and the only account of an ossifying fibroma causing a large craniofacial defect and significant orbital displacement. A local literature search suggests that this is the first account of a frontoethmoid sinus ossifying fibroma in the Philippines. The atypical presentation of the mass posed several surgical challenges, including difficult wide extirpation of the mass, preservation of the brain and eye and formidable reconstruction of the large defect.

The ossifying fibroma appeared in the CT scan as a single large, irregularly shaped multicystic heterogenous entity causing surrounding osteolytic changes. But on MRI, it appeared as a round, well encapsulated cystic mass surrounded by an expansile osteolytic mucocele that may have originated from an obstruction of the frontal recess. There are only a few reports of mucoceles caused by ossifying fibromas^{4,5} and this case is the first to show its formation in the frontal sinus.

The high recurrence rate of ossifying fibromas necessitates complete excision of the mass including involved orbital structures, frontal bone and dura. Although this radical extirpation can lead to debilitating complications like blindness, brain herniation, bleeding, cerebrospinal fluid leak and intracranial infections, the latter MRI findings of mucocele formation led to a more conservative surgical approach. This experience emphasizes the need for sufficient diagnostic imaging.

Orbital exenteration is an indicated treatment for malignant tumors, but there are indications for exenteration in benign orbital disease, including uncontrollable pain, blindness, cosmetic disfiguration and tendency of infiltration and malignant transformation.⁶ In this case, exenteration could address the possibility of orbital infiltration and cosmetically unacceptable proptosis but would also cause iatrogenic blindness. Ultimately, the orbit was preserved despite its inferior displacement because ophthalmological evaluation showed an intact globe, optic nerve and extraocular muscles.

There are no reports known to us of an ossifying fibroma causing direct intradural extension, although excision of a frontal sinus ossifying fibroma can cause iatrogenic CSF leak.⁷ Mucoceles associated with

obstructing paranasal sinus lesions have also been shown to cause dural defects and intradural expansion.⁸ This case showed that careful extirpation and thorough intraoperative neurosurgical evaluation can justify a conservative approach to the management of frontal bone defects.

Calvarial bone grafts have been used to reconstruct defects of the anterior cranial fossa and orbit to obtain satisfactory aesthetic and functional results with minimal complications.⁹ Abdominal fat has been used as a safe and practical orbital implant following ocular evisceration despite its gradual volume reduction over time.¹⁰ Temporalis muscle flaps have also been utilized in reconstruction of facial defects after excision of ethmoid tumors.¹¹ This case provides evidence that these autologous grafts can safely be used to reconstruct large craniofacial defects.

Currently the patient is well and free of recurrence. The proptosis has been markedly reduced allowing her to wear sunglasses to protect and conceal her left eye. A second stage orbital reconstruction is planned after six months to allow the orbit to assume a definite position after adequate wound healing, fat resorption and contraction.

In summary, a large ossifying fibroma of the frontoethmoid sinus is a rare condition appearing as an externally enlarging mass that can cause significantly more internal destruction while asymptomatic but gradual mucocele formation, bony erosions, brain involvement and orbital displacement put the patient in significant peril. CT scans and MRI are vital in accurate diagnosis and surgical planning. A multidisciplinary approach to complete surgical resection is needed for successful excision and functional orbital preservation. The use of autologous reconstructive methods such as calvarial bone graft, abdominal fat and temporalis muscle flaps were effective, safe and practical options in this case.

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