



Optic Nerve Compression Caused by Sphenoid Mucocele Secondary to a Fibrous Dysplasia: Case Report and Review of the Literature

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Abstract

We present an extremely infrequent case of a patient suffering from visual acuity loss due to optic nerve compression caused by sphenoid mucocele secondary to a fibrous dysplasia.

We present a 29-year-old man reporting a gradual loss of the visual acuity in the left eye during the last five days. The complete study, including MRI, revealed an extended fibrous dysplasia involving the right sphenoid and associated to a retention mucocele causing compression to the left optic nerve on its cisternal trajectory.

The patient underwent a left transnasal endoscopic sphenoidotomy with abundant drainage of the mucocele by marsupialization and creation of a new drainage pathway and preservation of the epithelium.

One month later, ophthalmologic examination revealed an important improvement on the visual acuity of his left eye.

Fibrous dysplasia accounts for 2% to 5% of all bone tumors. Mucoceles are benign masses that may provoke serious complications due to anatomical relationships when they grow. The apparition of both conditions associated is very rare.

Acknowledging the connection between the two conditions to get an accurate diagnosis is crucial. An early surgical procedure improves the functional prognosis of the patient.

Keywords: Sphenoid mucocele; Fibrous dysplasia; Optic nerve compression

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Abbreviations

MRI: Magnetic Resonance Imaging; RAPD: Relative Afferent Pupillary Defect; OCT: Optical Coherence Tomography; CT-Scan: Computed Tomography Scan

Introduction

We present the case of a 29-year-old male suffering from a loss of the visual acuity in the left eye due to the compression of the optic nerve caused by a sphenoid mucocele secondary to a fibrous dysplasia. This is a very infrequent case reported in the literature.

Fibrous Dysplasia (FD) is an infrequent noncancerous and slowly progressive disease consisting on normal bone marrow being replaced by immature fibro-osseous tissue, with a gradual expansion beyond its bony margins than can cause concomitant displacement of the surrounding soft tissue structures [1-3]. It accounts for 2% to 5% of bone tumors.

It can involve single bone (monostotic, 70% of cases) [1], or multiple bones (polyostotic). The monostotic form is less serious than the polyostotic form [1], and it appears more frequently in the jaws. It is called craniofacial FD when it affects the adjacent craniofacial bones [3].

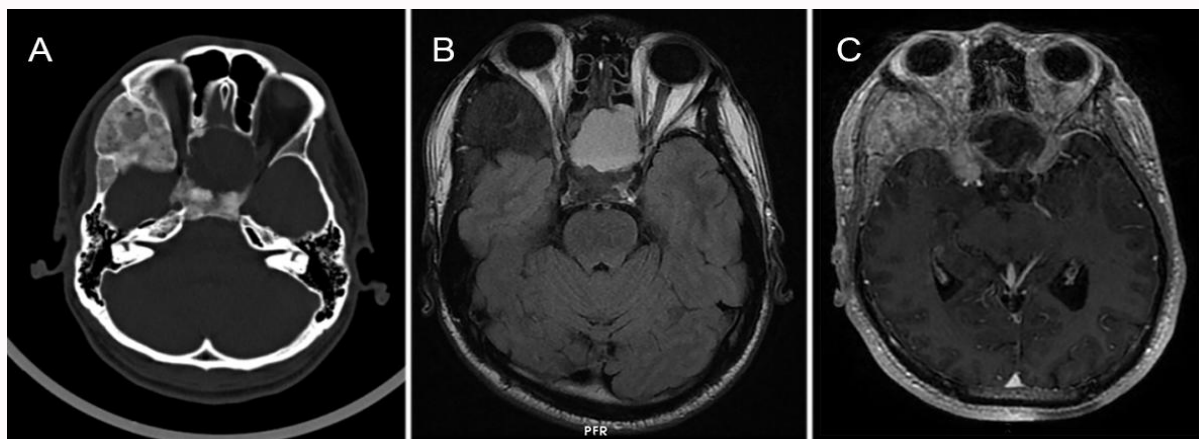


Figure 1A: CT image showing a bone diffuse enlargement of the major and minor wings of the right sphenoid, pterygoid process and body of the sphenoid, involving all the sphenoidal sinus.

Figure 1B: MRI showing the FLAIR sequence. The sphenoid sinus is substituted and enlarged by a liquid-mucoid cyst showing high-intensity.

Figure 1C: MRI T1- weighted image showing that the cyst has a subtle rim enhancement with gadolinium.

On the other hand, mucoceles are benign masses that, in some cases, may provoke serious complications due to anatomical relationships when they grow and they are filled with mucous and lined by epithelium. Around 1% to 2% of all paranasal sinus mucoceles are located in the sphenoid sinus [4], while most mucoceles are found in the frontal and/or the ethmoidal sinuses [5,6]. FD associated with sphenoid mucocele is a very rare condition [7], especially if it is associated with a compression of the optic nerve.

Case Presentation

A 29-year-old man arrived in the emergency room reporting a gradual loss of the visual acuity in the left eye during the last five days. The ophthalmologic examination revealed a visual acuity of 1.0 for the right eye but with left eye he was only able to perceive hand movements and he had a Relative Afferent Pupillary Defect (RAPD). The Optical Coherence Tomography (OCT) and the retinogram were normal. Visual evoked potentials did not have any response, so severe neuropathy was evinced. Physical examination showed chronic deformity on the right frontotemporal side.

The Computed Tomography Scan (CT-scan) (Figure 1A), and Magnetic Resonance Imaging (MRI) study showed a big bone diffuse enlargement on the major and minor wings of the right sphenoid, a pterygoid process and the body of the sphenoid, affecting all the sphenoidal sinus.

Furthermore, the sphenoid sinus is substituted and enlarged by a liquid-mucoid cyst, showing high-intensity on FLAIR (Figure 1B), and T2-weighted images and a delicate rim enhancement on the T1-weighted image with gadolinium (Figure 1C).

This retention cyst is affecting all the extra orbital trajectory of the left optic nerve, which seems to be thinner than usual.

The bone enlargement also causes facial asymmetry and proptosis of the right eye. It seems that the right optic nerve is also compressed on an intracanalicular and intracranial trajectory.

The MRI revealed an extended fibrous dysplasia involving the right sphenoid and associated to a retention mucocele causing compression to the left optic nerve on its cisternal trajectory.

The patient underwent a left transnasal endoscopic sphenoidotomy

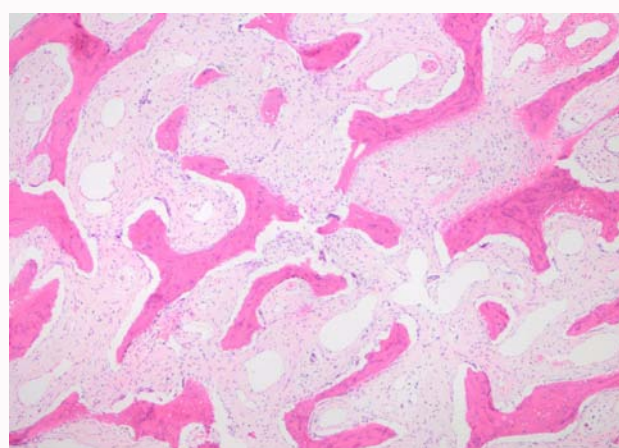


Figure 2: HE x40. The anatomic pathology study reveals irregular curvilinear discontinuous trabeculae of woven bone surrounded by a fibrous stroma, moderately cellular (without atypia or mitotic activity), which contains scattered blood vessels. The bone lacks prominent osteoblastic rimming, but osteoclasts may be numerous.

Our case contains areas of haemorrhage associated with multinucleated osteoclast-type giant cells mimicking giant cell reparative granuloma.

with abundant drainage of the mucocele by marsupialization and creation of a new drainage pathway and preservation of the epithelium [7]. The biopsy of the bone was done and the histopathological report confirmed a fibrous dysplasia (Figure 2). The patient was administered antibiotherapy since his admission.

One month after the surgery, the ophthalmologic examination revealed an improvement of the visual acuity of 0.6 in the left eye (previously only hand movements). The visual acuity in the right eye was preserved (1.0).

Two months after the surgery, the patient went through an endocrinological evaluation, which reported a normal functioning of the hypophysis. Postoperative images show the evacuation of the mucocele of the sphenoid sinus with no evidence of a left optic nerve compression (Figure 3).

Discussion

The loss of visual acuity due to an optic nerve compression caused

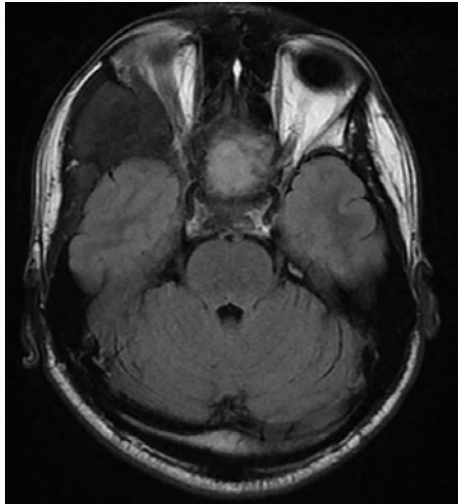


Figure 3: Postoperative FLAIR MRI sequence showing the complete evacuation of the mucocele in the sphenoid sinus. It also shows an improvement in the left optic nerve compression.

by a sphenoid mucocele secondary to a FD is extremely infrequent.

We performed a left endoscopic sphenoidotomy and drainage of the mucocele cyst, with a good visual outcome in the long term.

On the other hand, in this case, the MRI and CT scan images show that the FD is causing the compression of the right optic nerve. A conservative treatment is preferred for FD, unless there are neurological symptoms or serious deformity present [2]. The visual acuity loss due to an optic nerve compression has been described in FD. In these cases, optic nerve decompression is recommended [2]. However, several studies have shown that there was no correlation between the radiological compression and visual acuity [8,9]. In addition, prophylactic optic nerve decompression in FD has shown poor visual outcomes. Nowadays, literature is against the prophylactic decompression [10].

Conclusion

The visual acuity loss due to an optic nerve compression caused by a mucocele cyst secondary to a FD is a rare condition. It is

important to consider it when treating a patient with visual acuity loss and FD in order to get an accurate diagnosis. A fast and aggressive treatment from the beginning of the symptoms, consisting on surgery (endoscopic sinus approach with cyst drainage) is strongly recommended to improve the functional prognosis.

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