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Case Report

Rhinogenic Optic Atrophy - A Misdiagnosed and Misunderstood Cause of Preventable Blindness a Case Report and Review of Literature

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Abstract

Optic neuropathy caused by compression of the optic tract or chiasm due to mucoceles of the sphenoid sinus is called Rhinogenic optic atrophy. It is difficult to prognosticate owing to the poor incidence as well as delay in detection and treatment. The delay ensured in diagnosis and treatment ensures uniformly bad prognosis for vision post decompression. There are very few reports of Rhinogenic optic neuropathy and its recognition as a treatable entity vis-à-vis vision salvage is poorly recognized. We present a case report along with a review of the scanty literature present.

Keywords: *Rhinogenic Optic Neuropathy; Sphenoid Sinus Mucocele; Atypical Optic Neuropathy*

Introduction

Mucoceles of sphenoid sinus are rare and constitutes 1% of all paranasal sinus mucoceles [1,2]. It usually results from long standing sinusitis and closure of the sinus ostia. The expanding mucocele leads to osteoclastic absorption of the surrounding bony architecture and pressure effect on the adjacent neurovascular structures. Presentation of sphenoid sinus mucocele is variable depending upon the neurovascular structures involved. The optic neuropathy developing from these mucoceles has been termed rhinogenic optic neuropathy and once the patient develops visual disturbances, it should be treated as an emergency. The visual prognosis depends upon the visual acuity at presentation, size and location mucocele, and the duration between presentation and surgery [3-5]. Prognosis is poorer in cases where there is profound visual impairment at the time of presentation and delay in surgery.

We report a rare case of rhinogenic optic neuropathy who presented with longstanding right sided visual impairment. This patient presented to us after 6 months of progressive visual loss and was taken up for endoscopic decompression of the mucocele. Post-surgery, he improved well and was discharged with near normal vision. We hereby present a review of this scanty literature and compare our findings with them.

Case Report

A 54-year-old male presented to the outpatient clinic with progressive bifrontal headache for a year associated with gradual visual loss in the right eye for 6 months. The visual loss was initially in the lateral aspect of the field making him bump into people while walking on a crowded street and eventually progressed to almost complete loss of vision. The visual loss wasn't associated with difficulty in movement

of the eyeball, ptosis, seizures or hypothalamic disturbances. The left eye was completely normal. An MRI of the brain showed a large mass in the sphenoid sinus especially on the right aspect extending upto the mouth of the orbit compressing the optic nerve. The lesion was hyperintense on T1 and hypointense on T2 with no enhancement on contrast expanding and thinning the sphenoid sinus walls and compressing the optic nerve as mentioned above (Figure 1). Based on the presentation and radiological findings, a diagnosis of a sphenoid mucocele was made and the patient was counselled for surgery. He was taken up for a trans-nasal decompression of the mucocele under endoscopic guidance. The sphenoidal ostium was fibrosed and closed by the mucocele (Figure 2a) and was dissected to reveal the lesion inside the sinus cavity (Figure 2b). complete excision showed the exposed dura of the diaphragma sellae (Figure 2c) as the end point of the surgery. The cavity was then packed with gelfoam and closed. Post operatively his vision improved significantly. His visual acuity and fields pre- and post-surgery are documented in figure 3, which shows improvement not only in lateral field vision but also in acuity. The post-operative scan also showed good decompression (Figure 4). The patient was discharged to review in the outpatient clinic completely rid of his unusual ailment.

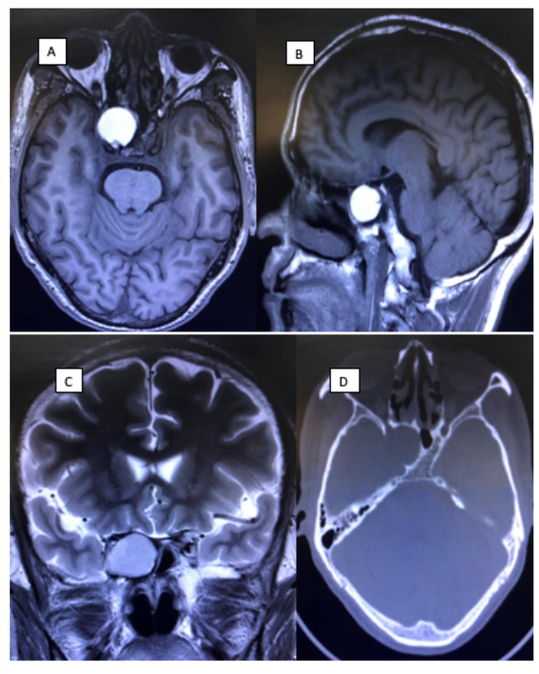

Figure 1: *Pre-operative MRI of the brain (A) T1 axial image, (B) T1 sagittal image, (C) T2 coronal image and (D) CT bone cuts in axial plane showing a large lesion in the right side of the sphenoid sinus compressing the mouth of the orbit on the right side.*

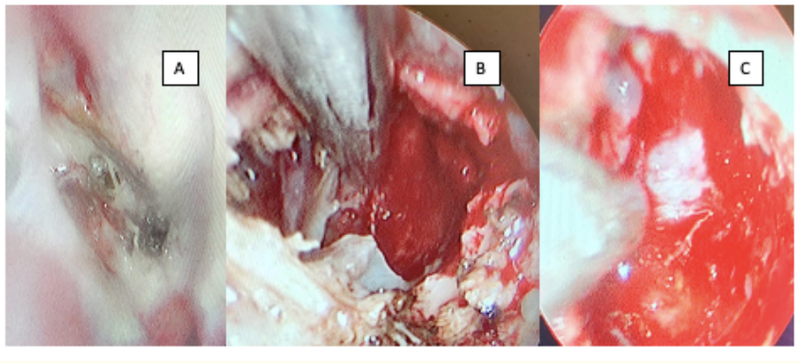

Figure 2: *Intraoperative pictures of the mucocele. Figure (A) shows the sphenoid ostium closed and fibrosed due to the mucocele, Figure (B) shows the excision of the mucocele in the sphenoid sinus, and Figure (C) shows the dura seen after surgery signifying adequate decompression.*

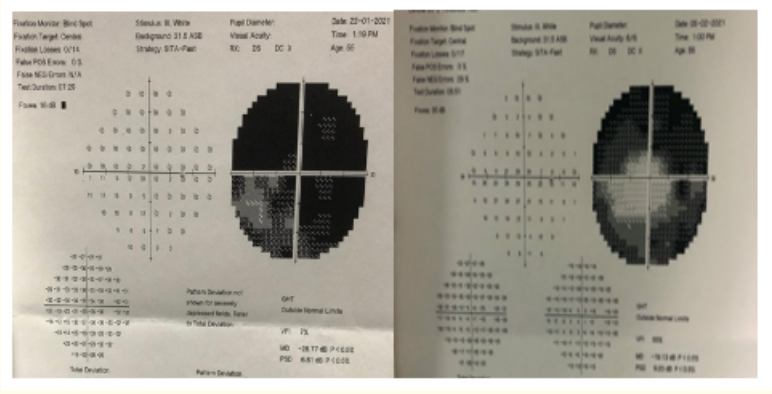

Figure 3: *Visual field charting pre- and post-surgery showing improvement in vision after decompression of the mucocele.*

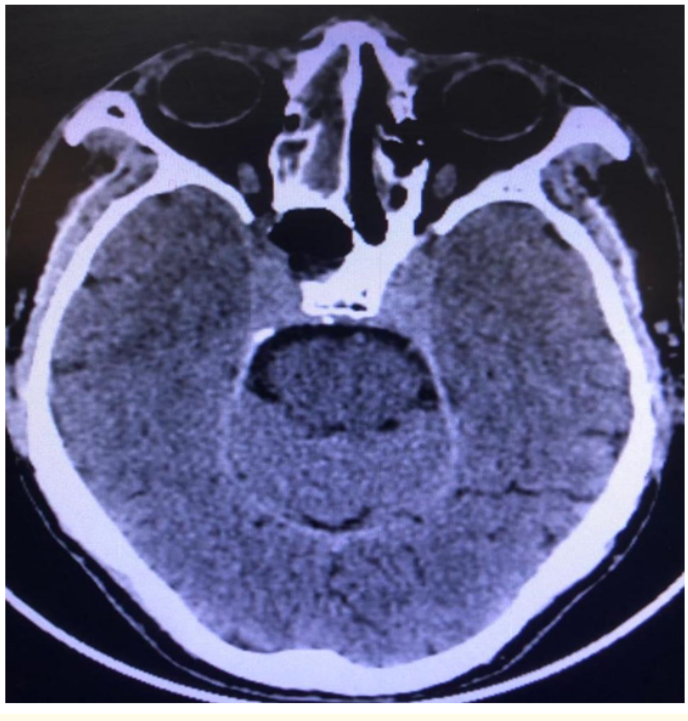

Figure 4: *Post-operative CT scan of the brain showing decompression of the sphenoid sinus and the mouth of the right orbit freeing the optic nerve.*

Discussion

Rhinogenic optic neuropathy is a rare differential diagnosis of optic neuritis. Mucoceles are cystic, respiratory epithelium lined structures which have ability to cause bone destruction within paranasal sinuses. Sphenoidal mucocele is very rare accounting for around 1% of all paranasal sinus mucoceles [1,2]. The spheno-ethmoidal cells are in close relation with the sphenoid sinus, optic nerve and internal

carotid artery. Because of this close proximity, the optic nerve may get involved in several ways. Direct spread of the sinus infection is the most common mode; causing an infective optic neuritis [6]. The cytokines released during the infective process stimulate fibroblasts to release prostaglandins and collagens which in turn stimulate bone destructions causing further expansion of the mucocele. This silent expansion of the mucocele may ultimately lead to compression of the optic nerve [7]. The released cytokines can also cause secondary inflammatory occlusive vasculitis and optic neuritis [8].

patients usually present with visual loss with or without motility disturbances. Afferent pupillary defect with visual field loss is usually seen. The classic radiological sign is appearance of large, distorted sinus with bone defect and compression of the optic nerve.

Rhinogenic optic neuropathy should always be considered as an neurosurgical emergency. Visual prognosis is extremely guarded and entirely depends upon the pre-operative vision and duration of the disease [3-5]. Prognosis is very poor in cases where visual loss is profound (hand movement, perception of light, no perception of light). It has been reported that visual prognosis is poorer if surgery is delayed for more than 6 - 10 days after vision loss and if optic atrophy has settled in [5]. McCarthy and Frenkel reported diminution in final visual acuity of 64% of their study subjects with sphenoid sinus mucocele [9]. They stated that the cause was pressure effect on optic nerve and/or central retinal artery and no improvement could be achieved in more than 50% of the cases, even after surgical intervention. The role of pre or post-operative steroid treatment in cases of rhinogenic optic neuropathy is a matter of debate, however we gave a one month course of post-operative oral steroid to our patient [4,10,11].

Fujimoto., *et al*. [12] in their study on optic nerve blindness due to paranasal diseases included 7 patients with no light perception vision. All underwent endonasal surgery within 4 days of onset of decreased vision. Five of the 7 patients had an increase in their final vision to 20/200 or better, however 2 patients didn't show any improvement in vision even after early surgery.

The study done by Nakaya., *et al*. [10] included 2 patients (out of total 38 patients) of rhinogenic optic neuropathy with no light perception vision with surgical delay of 4 and 16 days respectively. Both the patients failed to show any improvement.

Selvakumar and colleagues [13] reported a case of rhinogenic optic neuropathy, wherein patient had a 2 weeks history of vision loss in both the eyes. The delay between vision loss and FESS surgery was 16 days and there was no improvement in vision in one of the eyes even after surgery. Siritho., *et al*. [11] reported a case where the patient was misdiagnosed as optic neuropathy and there was a delay of one and a half month between visual loss and surgery. There was no improvement in the vision after surgery.

Otsuka., *et al*. [4] reported a similar case as ours, where the patient had sphenoid sinus mucocele with no perception of light. There was no delay between visual loss and surgery and the patient's vision improved to 0.3 Log Mar units in the post-operative period. They concluded that preoperative visual acuity should be considered as the most important predictive factor for postoperative visual acuity improvement. However, an improvement in visual acuity could be expected even in cases without light perception. A detailed comparative review of literature has been provided in table 1.

Parameters	Fugimoto., *et al.* (1999) [12]	Nakaya., *et al.* (2011) [10]	Siritho., *et al.* (2018) [11]	Selvakumar., *et al.* (2014) [13]	Otsuka., *et al.* (2019) [4]	Bose., *et al.* (2021)
Sample size(N)	NPL eyes-7	NPL eyes- 2	1	1	NPL eyes-2	1
Age/Sex	Mean age-51.42+/-10.75 years M:F=2:5	1. 74 Y/M 2. 76 Y/F	59 Y/M	17 Y/M	1. 64 Y/F 2. 76 Y/F	48 Y/F
Optic disc appearance	No signs of optic atrophy	1. Abnormal fundus 2. Normal fundus	Normal	Subtle temporal pallor	NA	WNL
Visual fields	NA	NA	NA	NA	NA	Normal for other eye
Cause of rhinogenic optic neuropathy	Ethmoidal cyst 3 Spheno-ethmoid cyst-2 Sphenoid cyst-1 Ethmoid sinusitis 1	Spheno-ethmoidal mucocele	Sphenoid mucocele	Spheno-ethmoidal mucocele	Spheno-ethmoidal mucocele and pyocele	Sphenoid sinus mucocele

Surgical delay	Within 4 days	1. 4 days 2. 16 days	6 weeks	16 days	1. 0 Days 2. 4 Days	23 days
Surgical procedure	Endonasal sinusotomy -3 Endonasal ethmoidectomy/ sphenoidotomy with drainage 4	FESS with drainage and preservation of mucosa facing skull base	Posterior spheno-ethmoidectomy with drainage	Right transethmo- sphenoidal drainage of the mucocele	Endoscopic sinus surgery	Functional endoscopic sinus surgery (FESS) with drainage of mucocele
Follow up	1-120 months	6 months	10 days	2 months	Not mentioned	18 months
Post operative vision at final follow up	5 patients >/= 20/200 2 patients- NPL	NPL	NPL	NPL	Both NPL	20/400
HVF after surgery	NA	NA	NA	NA	NA	Central scotoma
Optical coherence tomography for RNFL and GCL	NA	NA	NA	NA	NA	RNFL thinning in temporal quadrant, GCL thinning in all quadrants

Table 1: *Review of literature of previously reported cases of rhinogenic optic neuropathy presenting with no perception of light vision. NPL: No Perception of Light; M-Male; F-Female; Y-Years; NA-Not Available; HVF- Humphrey Visual Field; RNFL: Retinal Nerve Fibre Layer; GCL-Ganglion Cell Layer.*

Conclusion

Rhinogenic optic neuropathy is an important differential diagnosis of optic neuritis. Delay in diagnosis and management can lead to permanent vision loss. Although timely management can prevent irreversible loss of vision and salvage useful vision to some extent; delayed surgery too in rare circumstances can expect visual recovery.

Prior Publication

Nil.

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Nil.

Conflicts of Interest

Nil.

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