

# Partial Visual Recovery after Delayed Surgery in a Case of Rhinogenic Optic Neuropathy: Report of a Case and Review of Literature

## Case Report

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### Abstract

Rhinogenic optic neuropathy is a clinical term used for optic neuritis or neuropathy caused by paranasal cysts or mucocele. Optic neuropathy associated with sphenoid sinus mucocele is usually associated with poor prognosis. The prognosis in all these cases depends upon visual acuity at presentation and duration of disease. Delay in surgery in all such cases leads to extremely poor visual outcome. We herewith report a rare case of rhinogenic optic neuropathy that presented with complete loss of vision and had partial recovery of vision even after surgery being delayed for almost 3 weeks.

**Keywords:** Rhinogenic optic neuropathy; Sphenoid sinus mucocele; Atypical optic neuropathy

## Introduction

Mucocele of sphenoid sinus is 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 herewith report a rare case of rhinogenic optic neuropathy who presented with unilateral no light perception vision, was operated after 23 days from the commencement of vision loss and showed significant improvement in vision to finger counting at three meter at the last follow up. To the best of our knowledge there are limited case reports of rhinogenic optic neuropathy presenting with complete loss of vision showing partial visual recovery even after delayed surgery. We have also discussed the optical coherence tomography findings which have not been reported in the literature for these cases.

## Case Report

A 48 years old female presented with complains of right eye gradually progressive painful loss of vision for the past 2 weeks. The visual disturbance was preceded by episode of headache, nausea and

vomiting. Patient had history of recurrent upper respiratory tract infection and was a known case of diabetes mellitus, hypertension and hypothyroidism.

On examination right eye had no perception of light while it was 20/20 in the left eye. The right eye had a relative afferent papillary defect (RAPD) with mild temporal tenderness. Extra-ocular movements were full. Rest of the anterior and posterior segment examination was within normal limits. Left eye was otherwise normal. MRI of brain and orbits showed features suggestive of large expansile cystic lesion in the sphenoid sinus. The lesion was hyperintense on T1 and T2 weighted sequences and exhibited rim enhancement (Figure 1 A). It was compressing over the intra-canalicular part of the right optic nerve with abnormal contrast enhancement in this segment (Figure 1 B). The findings were suggestive of sphenoid sinus mucocoele with right sided rhinogenic optic neuropathy.

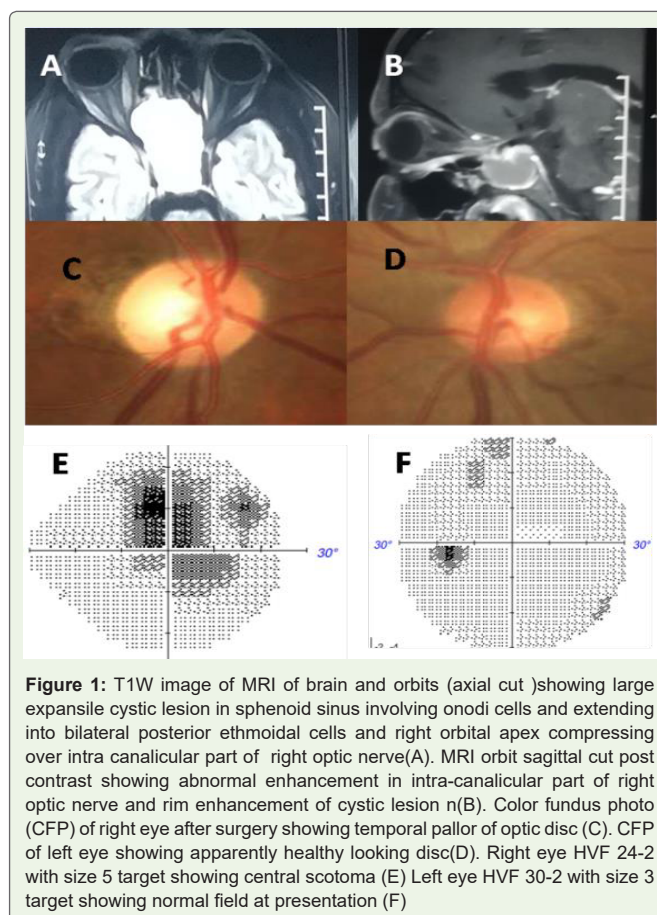
The patient was immediately referred to an ENT surgeon, however he underwent functional endoscopic sinus surgery (FESS) and drainage of the mucocoele after 8 days causing a total delay of 23 days between onset of visual loss and surgery. The patient reported back to us after a week of surgery and the vision noted was counting finger at one meter. Patient was given a trial of oral steroids for a month to take care of any residual inflammation from the disease process.

After three months the vision improved to counting fingers at three meters. Temporal pallor of the optic disc had settled in by this time (Figure 1 C), optic disc of the other eye was however normal (Figure 1 D). The patient was able to perform perimetry with large target which showed fixation scotoma (Figure 1 E). Humphrey visual field of the left eye was however normal both at the presentation and at the final visit (Figure 1 F).

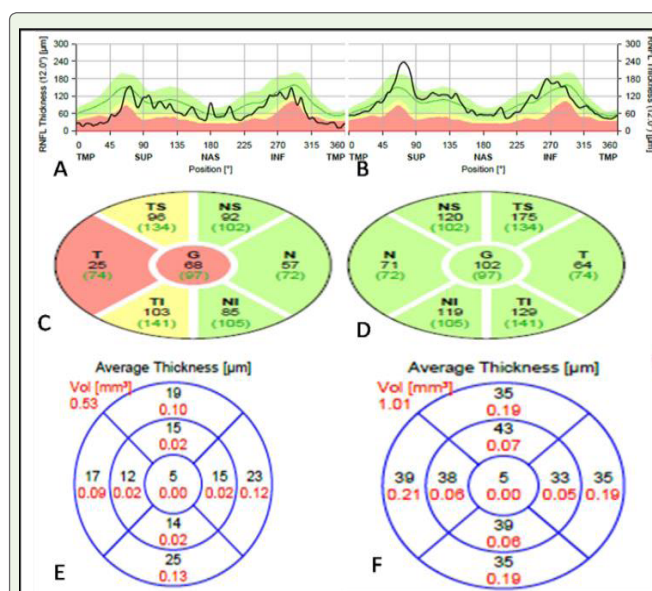
Right eye optical coherence tomography(OCT) (Spectralis OCT platform, Heidelberg engineering, Heidelberg Germany) for analysis of retinal nerve fiber layer (RNFL) and ganglion cell layer (GCL) was performed at this visit which showed significant RNFL thinning in temporal quadrants and GCL thinning in all quadrants in the right eye as compared to left (Figure 2).

## Discussion

Rhinogenic optic neuropathy is a rare differential diagnosis of optic neuritis. Mucocoeles are cystic, respiratory epithelium lined structures which have ability to cause bone destruction within paranasal sinuses. Sphenoidal mucocoele is very rare accounting for around 1% of all paranasal sinus mucocoeles [1,2]. The sphenoid-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 [3]. 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 mucocoele. This silent expansion of the mucocoele may ultimately lead to compression of the optic nerve [7]. The released cytokines can also cause secondary inflammatory occlusive vasculitis and optic neuritis [8].



**Figure 1:** T1W image of MRI of brain and orbits (axial cut) showing large expansile cystic lesion in sphenoid sinus involving onodi cells and extending into bilateral posterior ethmoidal cells and right orbital apex compressing over intra canalicular part of right optic nerve (A). MRI orbit sagittal cut post contrast showing abnormal enhancement in intra-canalicular part of right optic nerve and rim enhancement of cystic lesion (B). Color fundus photo (CFP) of right eye after surgery showing temporal pallor of optic disc (C). CFP of left eye showing apparently healthy looking disc (D). Right eye HVF 24-2 with size 5 target showing central scotoma (E) Left eye HVF 30-2 with size 3 target showing normal field at presentation (F)



**Figure 2:** OCT retinal nerve fiber layer (RNFL) analysis showing thinning of RNFL in temporal quadrants in right eye (A,C). OCT RNFL analysis of left eye showing normal thickness (B,D), OCT ganglion cell layer analysis map of right eye (E) showing thinning of ganglion cell layer in all outer and inner circles compared to left eye (F).

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 ophthalmic/rhinological emergency. Visual prognosis is extremely guarded and entirely depends upon the pre-operative vision and duration of the disease [4-6]. 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 mucocoele [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].

In the present case there was a delay of 23 days between commencement of diminution of vision and surgery. However the vision improved from no light perception to 20/400 at 18 months of follow up period.

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 colleague [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 mucocoele 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.

Detailed comparative review of literature has been provided in (Table 1).

**Table 1:** Review of literature of previously reported cases of rhinogenic optic neuropathy presenting with no perception of light vision

Parameters	Fujimoto et al (1999) [12]	Nakaya et al (2011) [10]	Siritho et al (2018) [11]	Selvakumar et al (2014) [13]	Otsuka et al (2019) [4]	Present case
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	Ethemoidal cyst-3 Spheno-ethemoidal cyst-2 Sphenoid cyst-1 Ethmoid sinusitis-1	Spheno-ethemoidal mucocoele	Sphenoid mucocoele	Spheno-ethemoidal mucocoele	Spheno-ethemoidal mucocoele and pyocoele	Sphenoid sinus mucocoele
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/ sphenoidectomy with drainage-4	FESS with drainage and preservation of mucosa facing skull base	Posterior spheno-ethmoidectomy with drainage	right transthemio-sphenoidal drainage of the mucocoele	Endoscopic sinus surgery	Functional endoscopic sinus surgery (FESS)with drainage of mucocoele
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	1. 0.3 LogMar 2. 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

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

To the best of our knowledge there are no reports on OCT findings in cases of rhinogenic optic neuropathy. Though we have missed the preoperative OCT examination, we strongly feel that preoperative OCT RNFL and GCL analysis could predict visual prognosis after surgery.

## 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.

## References

1. Prepageran N, Subramaniam KN, Krishnan GG, Raman R (2004) Ocular presentation of sphenoid mucocoele. *Orbit* 23: 45-47.
2. Levy J, Monos T, Puterman M (2005) Bilateral consecutive blindness due to sphenoid sinus mucocoele with unilateral partial recovery. *Can J Ophthalmol* 40: 506-508.
3. Dale BA, Mackenzie IJ (1989) The complications of sphenoid sinusitis. *J Laryngol Otol* 97: 661-670.
4. Otsuka M, Yunoki T, Ozaki H, Tachino H, Takakura H, et al. (2019) Prognostic Factors for Postoperative Visual Acuity in Patients with Rhinogenic Optic Neuropathy. *J Ophthalmol* 2019: 3417425.
5. Moriyama H, Hesaka H, Tachibana T, Honda Y (1992) Mucocoeles of ethmoid and sphenoid sinus with visual disturbance. *Arch Otolaryngol Head Neck Surg* 118: 142-146.
6. Yumoto E, Hyodo M, Kawakita S, Aibara R (1997) Effect of sinus surgery on visual disturbance caused by sphenoid mucocoeles. *Am J Rhinol* 11: 337-343.
7. Newton N, Jr., Baratham G, Sinniah R, Lim A (1989) Bilateral compressive optic neuropathy secondary to bilateral sphenoid mucocoeles. *Ophthalmologica* 198: 13-19.
8. Moorman CM, Anslow P, Elston JS (1999) Is sphenoid sinus opacity significant in patients with optic neuritis? *Eye* 13: 76-82.
9. McCarthy WL, Frenkel M (1972) Visual loss as the only symptom of sphenoid sinus mucocoele. *Am J Ophthalmol*. 74: 1134-1140.
10. Nakaya K, Oshima T, Kudo T, Ikeda R, Hidaka H, et al. (2011) Outcomes of surgically treated rhinogenic optic neuropathy. *Acta Otolaryngol* 131: 833-839.
11. Siritho S, Tantiniorn W, Wichiwaniwate P, Pongpirul K (2018) Rhinogenic Optic Neuritis Caused by Sphenoid Mucocoele with Sinusitis. *Case Rep Radiol* 2018: 8302415.
12. Fujimoto N, Adachi-Usami E, Saito E, Nagata H (1999) Optic nerve blindness due to paranasal sinus disease. *Ophthalmologica* 213: 262-264.
13. Selvakumar A, Mahalaxmi B, Ananth V, Gautam C (2014) Sphenoidal mucocoele causing bilateral optic neuropathy and ophthalmoplegia. *Indian J Ophthalmol* 62: 515-517.