Coexistent Optic Neuritis and Isolated Acute Sphenoid Sinusitis

MG Rajiniganth, Usha R Kim, Mahesh Kumar

1Endoscopic Sinus, Orbit, Skull Base Specialist, Aravind Eye Hospital and Postgraduate Institute of Ophthalmology, Madurai Tamil Nadu, India
2Chief, Orbit, Oculoplasty and Oncology Clinic, Aravind Eye Hospital and Postgraduate Institute of Ophthalmology, Madurai Tamil Nadu, India
3Consultant, Neuro-ophthalmology Clinic, Aravind Eye Hospital and Postgraduate Institute of Ophthalmology, Madurai Tamil Nadu, India

Correspondence: MG Rajiniganth, Endoscopic Sinus, Orbit, Oculoplasty and Oncology Clinic, Aravind Eye Hospital and Postgraduate Institute of Ophthalmology, Madurai, Tamil Nadu, India, Phone: 91-452-5356100, Fax: 91-452-2530984, e-mail: mgrajini@gmail.com

Abstract
We report two cases of optic neuritis associated with isolated acute sphenoid sinusitis. This is the first report of isolated sphenoid sinusitis with dehiscent optic canal with exposed nerve presented with optic neuritis without any orbital inflammation. In both cases visual acuity deteriorated with steroid therapy, CT scan showed air fluid in the sphenoid sinus with dehiscent optic canal. Antibiotic, sphenoidotomy and later steroid course had a partial recovery in vision.

Keywords: Sphenoid sinus, optic canal, visual acuity.

INTRODUCTION
Optic neuritis describes an inflammation of the optic nerve caused by a wide variety of conditions; multiple sclerosis continues to be the most common. It is mainly a clinical diagnosis and they generally present with monocular visual deficits. RAPD is usually seen but the disk has a normal presentation as the inflammatory process is usually posterior to the optic disk. It is unusual for optic neuritis to coexist with sinus disorder and the simultaneous appearance of both diseases would invite etiological suspicion but management dilemma. We present very unusual cases of acute optic neuritis with associated isolated acute sphenoid sinusitis with exposed intracanalicular optic nerve due to dehiscent optic canal.

CASE 1
A 14-year-old girl presented with bilateral diminution of vision managed elsewhere with injection Methyl Prednisolone. The vision further deteriorated and she was referred to Neuro-ophthalmology clinic, Aravind Eye Hospital, Madurai for further management. On examination, the best corrected visual acuity was 6/60 in the right eye and 3/60 in the left eye. A relative afferent pupillary defect (RAPD) was present in the left side and other ocular examination was found to be normal. Confrontation visual field revealed a temporal field defect on the both sides and optic disk showed temporal pallor in both eyes. Visual acuity further deteriorated with IV steroid in the left eye to PL (Perception of Light) positive. Imaging of brain and orbit revealed a air fluid level in the sphenoid sinus and made them to get ENT consultation.

On specific ENT clinical evaluation she had complaints of postnasal discharge for the past 15 days since the onset of visual loss and associated ear pain in the both ear. Endoscopic evaluation revealed a mucopurulent discharge from the both sphenoid ethmoidal recess area along with congestion of nasopharynx. CT evaluation of sinuses showed air fluid level in both sides of sphenoid sinus. Left side of sphenoid sinus was found to be nondominant with dehiscent optic canal (Figs 1 and 2). She was started on injection Ceftriaxone, Gentamicin and considered for surgical drainage.

She underwent endoscopic transnasal sphenoileotomy under GA. Purulent discharge from the sphenoehtmoidal recess was sucked out, sphenoid ostium was blocked with granulation tissue which is removed. Ostium was widened with mushroom forceps and pus was sucked out. Same procedure was done in both sides. Left optic canal was found dehiscent and the nerve sheath was found to congested and thickened scalp vein set cannula was kept inside the
Patient was diagnosed as recurrent primary optic neuritis and treated with steroid. Visual acuity worsened further to PL positive and inaccurate projection of rays. CT brain showed air fluid level in the sphenoid sinus. On ENT evaluation he had complaints of headache (10 days, dull aching in the retro-orbital area) and postnasal discharge (Fig. 3). Nasal endoscopy revealed streak of mucopurulent discharge from the right sphenethmoidal recess area into nasopharynx, granulation tissue blocking the sphenoid ostium. Steroid was discontinued and IV antibiotic was started. The patient underwent sphenoidotomy under local anesthesia in the right side by endoscopic transnasal approach. Optic nerve was found exposed with dehiscent optic canal. The nerve found to be thickened and congested (Fig. 4). The patient was restarted with steroid and final visual acuity was 2/60.

DISCUSSION

The sphenoid sinuses are located in the center of the skull. Pneumatization of the sphenoid bone occurs during middle childhood and reaches its final form at the age of 12 to 15 years. The sinuses occupy a midline position in the coronal plane, and they are separated by an intersinus septum. The position of the sphenoid sinus septum varies greatly, and it can often be found well off the midline. They communicate with the superior nasal meatus by means of a small ostium of 0.5 to 4.0 mm, which is located disadvantageously 10 to 20 mm above the sinus floor. Several vital structures lie close to the sinus like the pituitary gland above, optic nerves, internal carotid arteries, and cavernous sinuses. Among these the understanding of relationship between the intracanalicular optic nerve and posterior group of sinuses is important.
Coexistent Optic Neuritis and Isolated Acute Sphenoid Sinusitis

There are four types of optic nerve relations with the posterior sinuses. Type I when the nerve runs along the side wall of the sphenoidal sinus without producing any indentation on the wall (76%); type II, the same as type I but causing indentation in the wall of the sinus (15%); type III when the nerve runs through the sphenoidal sinus (6%); and type IV when the nerve passes immediately adjacent to the sphenoidal sinus and the posterior ethmoid (3%). The type II and III optic nerve are more prone to get involved in coexistent sphenoid sinus pathology. A proper and methodical evaluation of imaging of sphenoid sinus will help to correlate with clinical presentation and wise management not only save vision but also life too.

Isolated sphenoid sinus diseases are not frequent and often the presentation is variable and nonspecific. The common specific presentations are headache, postnasal discharge and visual compromise. Because of variable signs and symptoms it presents to Physician, Neurologist (Headache), Ophthalmologist (visual compromise and diplopia). Usually imaging is done to brain or orbit and otolaryngologist assistance sought due to isolated sinus opacification in imaging. The otorhynologist must have a thorough knowledge of the anatomy, radiology and clinical spectrum of sphenoid sinus disease to evaluate and manage these patients properly. In our both cases it was treated primarily by ophthalmologist as optic neuritis and ENT assistance seeked in view of worsening visual symptoms and a positive imaging finding in sphenoid sinus.

The eye presentation of isolated sphenoid sinus disease are retro-orbital pain, diplopia and visual compromise which includes blurred vision and loss of visual acuity ranging from mild loss to total blindness. The incidence of visual loss is 12% of inflammatory cases, 60% of neoplastic cases. There are various theories for these visual compromise (a) direct spread of the sinus infection and inflammation to the optic nerve through dehiscent optic canal. (b) compressive optic neuropathy due to expansile or mass lesion in posterior ethmoid and sphenoid sinus; (c) vasculitis with thromboangitis of the optic nerve (d) bacteraemias resulting from the passage of infection through the mucosa of the sinus. In our case the visual loss is due to inflammation of optic nerve due to exposed nerve, in both cases the nerve was found thickened and congested.

Isolated acute sphenoid sinusitis and optic neuritis is a very rare combination and specifically with dehiscent optic canal with exposed optic nerve. In these combination both invite etiological suspicion on visual loss. Visual acuity in optic neuritis generally improves without treatment but high-dose intravenous methylprednisolone or oral prednisone demonstrated a more rapid return to normal vision, preventing recurrences, and reducing the incidence of multiple sclerosis (demyelinating disorder of optic nerve). Sphenoid sinus infection and subsequent inflammation is caused by insufficient clearing of mucus secondary to obstruction of the ostium. Isolated acute sphenoid sinusitis is often characterized by pain (recent onset, moderate to severe, aching or dull in quality, in the retro-orbital area, the occiput, or the vertex), nasal congestion and postnasal discharge. Evaluation of sphenoid sinus requires and nasal endoscopy and CT. Nasal endoscopy is an important clinical tool in the diagnosis of sphenoid sinus disease. Mucosal congestion, edema, purulent discharge are suggestive acute inflammation in endoscopy. CT is the standard investigation choice to diagnose sphenoid sinus disease and its complications. MRI is an essential adjunct mainly in sphenoid sinus lesion with intracranial complication. In the scan acute sinusitis appear as air fluid level with in the sinus cavity.

Complication of sphenoid sinusitis is rare but with devastating consequences. The orbital complication are orbital cellulitis, and orbital abscess. The intracranial complication are epidural abscess, cavernous sinus thrombosis and intracerebral abscess. The isolated optic neuritis without associated orbital cellulitis is very rare orbital complication of acute sphenoid sinusitis. It is usually associated with dehiscent optic canal with exposed intracanalicular segment of optic nerve to sphenoid sinus inflammation.

Acute sphenoid sinusitis is usually managed with antibiotic, decongestant and anti-inflammatory. Patients who present with severe symptoms may require Intravenous...
antibiotics. Patient with visual compromise is best managed similar to peripheral facial palsy in ASOM. Antibiotic with or without steroid and myringotomy in case of intact tympanic membrane is the standard treatment protocol in facial palsy in ASOM. Facial nerve decompression is controversial and nerve sheath is never incised in ASOM Cases.\textsuperscript{8,9} Similar principle of treatment is practiced in optic neuritis in acute isolated sphenoid sinusitis. If the optic canal is dehiscent, sphenoidotomy must be done to drain the pus and to remove the direct source of infection and inflammation to the exposed optic nerve. In all cases of retrobulbar optic neuritis sphenoid and posterior ethmoid sinuses pathology must be contemplated. The combined management of sinusitis and optic neuritis may prevent a permanent visual loss and good visual recovery.

CONCLUSION

Sphenoid sinus disease must be contemplated as a possible cause in all cases of retrobulbar optic neuritis. Imaging should be properly studied particularly the relationship between optic nerve and sphenoid and posterior ethmoid. The treatment of optic neuritis secondary to isolated acute sphenoid sinusitis with dehiscent optic canal should be antibiotics, anti-inflammatory, decongestant followed by sphenoidotomy to drain the pus and then steroid.

REFERENCES