

# Unilateral Optic Neuropathy due to a Sphenoid Mucocoele

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## ABSTRACT

Isolated sphenoid sinus mucocoeles are rare; whereas, benign lesions usually discovered incidentally. These may present with headache, diplopia, visual loss or ocular motor nerve palsies, due to slow expansion and mass effects. Prompt endoscopic removal is the key to prevent permanent sequelae. We present the case of a 63-year female, who presented with sudden loss of vision of the left eye, along with periorbital and retrobulbar pain for the last one month. She was found to have “no light perception” vision in the left eye and optic atrophy. Neuroimaging was performed, which revealed a sphenoid sinus mucocoele, impinging on the left optic nerve. Our ENT Department performed endoscopic sinus surgery and removed it, but her vision did not return at 6-month follow-up. Although very rare, sphenoid sinus mucocoeles should be considered in the differential diagnosis of any patient, who presents with sudden visual loss.

**Key Words:** Sphenoid sinus, Mucocoele, Optic neuropathy, Visual loss.

**How to cite this article:** Nadeem S, Akbar A, Zafar AN. Unilateral Optic Neuropathy due to a Sphenoid Mucocoele. *J Coll Physicians Surg Pak* 2021; **31(01)**:92-94.

## INTRODUCTION

Mucocoeles are benign, encapsulated, mucus filled, expansive paranasal sinus (PNS) masses lined by pseudostratified ciliated columnar epithelium. Isolated sphenoid sinus mucocoeles described by Berg for the first time in 1889, are the rarest of all PNS mucocoeles, with an incidence of only 1%. Less than 200 cases have been reported in literature worldwide so far.<sup>1-3</sup>

Symptoms depend on the anatomic location of the mucocoele, its direction of expansion and associated infection, if present. The most common presenting symptom is headache, followed by visual disturbances, diplopia, proptosis and rhinorrhea. Endoscopic sinus surgery is the preferred approach of treatment. Early diagnosis and treatment are the key to prevent irreversible damage to the optic nerve.<sup>1-3</sup>

We present the first instance of optic neuropathy due to a sphenoid sinus mucocoele to be reported in our country.

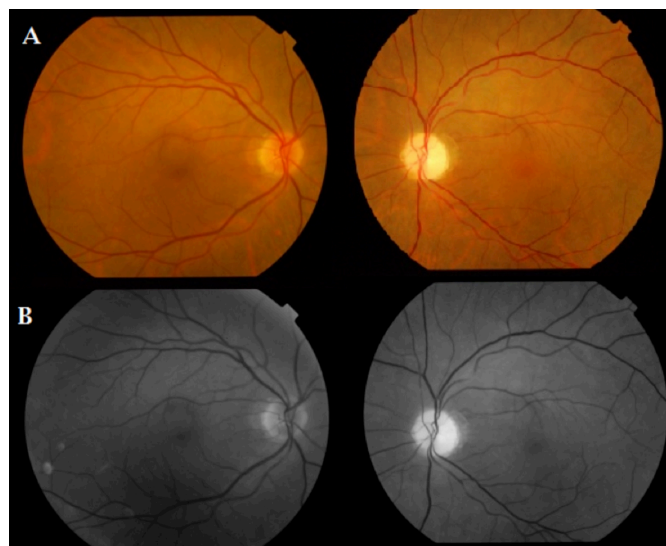
## CASE REPORT

A 63-year hypertensive female presented to us in the Eye Outpatient Department (OPD), with sudden visual loss in her left eye, accompanied by periorbital and retrobulbar pain for the last one month. She had undergone left phacoemulsification with an intraocular lens implant six months prior to presentation, at an outside facility.

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Received: July 11, 2019; Revised: October 05, 2019;  
Accepted: October 07, 2019  
DOI: <https://doi.org/10.29271/jcpsp.2021.01.92>

On examination, she had “no light perception” (NPL) in the left eye, quiet pseudophakia, along with a relative afferent pupillary defect (RAPD) and optic atrophy (Figure 1). The right eye was normal with a best corrected visual acuity of 6/9. The intraocular pressures were 17 mm Hg bilaterally. Both fundii had mild arteriovenous (AV) nicking and venous congestion was observed on the left. She was taking telmisartan-hydrochlorothiazide, 40 mg, once a day, for hypertension, escitalopram, 10 mg, for depression, and enteric coated aspirin, 75 mg, daily. She did not have any other systemic ailment or allergies.



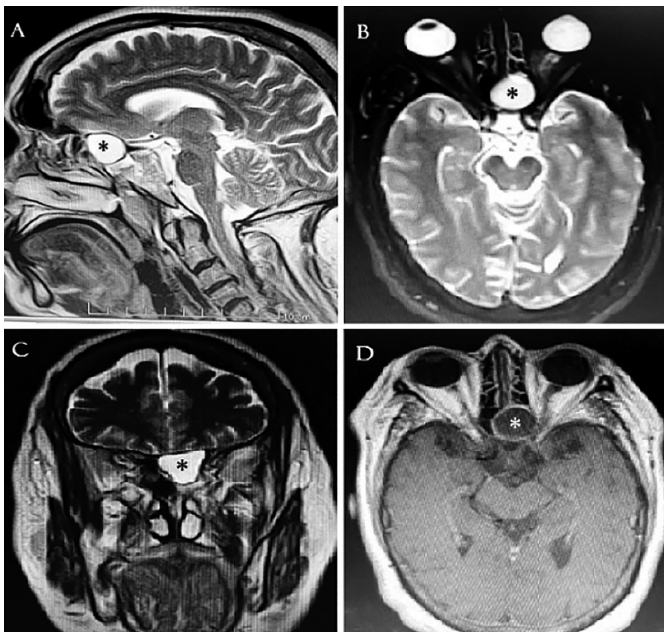
**Figure 1: (A) Colour fundus photographs showing left optic atrophy and venous congestion. (B) Red-free photographs.**

She was investigated thoroughly. Complete blood picture was normal with a hemoglobin of 13.8 g/dL. Liver function tests, serum urea, creatinine, serum lipid profile, and serum electrolytes were normal. Urine routine examination was normal as well.

Fasting plasma glucose was 4.2 mmol/L. Thyroid function tests were in normal range. The pituitary hormone levels were not raised. An initial CT scan of the PNS, orbit and brain was done which led to the diagnosis of a sphenoid mucocoele with optic nerve compression (Figure 2). Subsequently, a contrast-enhanced MRI of the orbit, PNS and brain was performed, which showed a 2.2 x 2 x 1.8 cm sphenoid mucocoele impinging on the left optic nerve, orbital apex and extending into the ethmoidal air cells (Figure 3).



**Figure 2:** CT scan orbit, paranasal sinuses (PNS) and brain showing sphenoid mucocoele (asterisk) extending into the orbital apex and ethmoid sinuses. (A) Sagittal view (B) Coronal view (C) Axial view.



**Figure 3:** MRI of orbit, paranasal sinuses (PNS) and brain [Contrast enhanced] showing an isolated sphenoid sinus mucocoele (asterisk), hyperintense on T2-weighted imaging and FLAIR, with fluid level and rim enhancement with gadolinium. (A) 35Sagittal T2W MRI showing the hyperintense mucocoele. (B) Axial T2W MRI showing optic nerve compression. (C) Coronal T2W MRI showing extension into ethmoidal air cells. (D) Axial T1W MRI showing cyst wall enhancement.

She was referred urgently to our ENT Department, where functional endoscopic sinus surgery (FESS) was performed to decompress the mucocoele. She was given systemic steroids for two weeks and was kept on multivitamin tablets, but her vision remains NPL at 6-month postoperatively.

## DISCUSSION

Sphenoid sinus mucocoeles or mucopyocoeles occur in response to multiple factors; mostly due to inflammatory causes like infection, sinusitis and allergies, or due to ostium or secretory gland obstruction as a result of mechanical insults like trauma, surgery, radiation, nasal polyps, or fungal masses. They are usually seen in the second to seventh decades of life. They slowly enlarge and expand along the path of least resistance causing an array of symptoms, although these may also be detected incidentally in asymptomatic individuals.<sup>1-3</sup>

Important structures in the vicinity of the sphenoid sinus tend to get affected by this expansion; these include most importantly the optic nerve, cranial nerves III, IV, V and VI, cavernous sinuses, internal carotid arteries, pituitary gland, optic chiasma, superior orbital fissure, and sphenopalatine nerve. The ethmoidal air cells and the nasopharynx are also common routes of invasion.<sup>3</sup>

The presenting features are worsening retro-orbital or frontal headaches (87%), visual loss which may be bilateral and severe (58%), diplopia due to involvement of cranial nerves III, IV, or VI (55%), and proptosis due to orbital invasion. Neuralgia due to trigeminal involvement, nasal symptoms like rhinorrhea (50%)<sup>1</sup>, hypoacusis, and pituitary dysfunction (3%) including permanent diabetes insipidus or panhypopituitarism (0.8%) have also been reported.<sup>2</sup>

The differential diagnosis of masses in this region includes chronic sphenoid sinusitis, fungal granuloma, pituitary adenoma, craniopharyngioma, glioma, meningioma, inverted papilloma, and nasopharyngeal carcinoma. MRI with contrast is needed for definitive diagnosis and for differentiation from other tumors.<sup>1-5</sup>

Optic neuropathy due to sphenoid sinus disease has various mechanisms; direct spread of inflammation leading to optic neuritis, compression, vascular compromise, occlusive vasculitis, osteomyelitis, orbital apex syndrome or presence of the onodi cell. Visual prognosis depends on the severity and duration of visual loss, and prompt therapy. Amaurosis due to sphenoid sinus disease is a rhinological emergency and urgent treatment is required to prevent irreversible optic nerve dysfunction.<sup>4,5</sup> In our patient, duration at presentation of one month, "no light perception" vision, and delayed diagnosis, all contributed to irreversible visual loss and no recovery whatsoever was seen, despite adequate surgical intervention.

The complications of sphenoid sinus mucocoeles that can arise are infection, mucopyocoeles, bony erosion, meningitis, cavernous sinus thrombosis, which should not be neglected.<sup>4,6</sup>

Medical treatment of sphenoid mucocoeles with antibiotics and steroids is effective in only 10% cases.<sup>7</sup> Endoscopic sinus surgery is considered the gold standard therapy; and is found to be very safe and effective. Other treatment approaches are trans-septal, trans-nasal, external or trans-nasal sphenoidectomy or intraoral transpalatal. Recurrence is rare.<sup>7</sup>

A high index of suspicion is required and a sphenoid sinus mucocoele should be considered in the differential diagnosis of all cases of sudden visual loss, which needs confirmation by neuroimaging. Early diagnosis and prompt surgical intervention are the key to visual recovery.

**PATIENT'S CONSENT:**

Informed consent was obtained from the patient to publish the data concerning this case.

**CONFLICT OF INTEREST:**

Authors declared no conflict of interest.

**AUTHORS' CONTRIBUTION:**

SN: Principal author, manuscript drafting, writing, editing, photography and patient follow-up.

AA: Performed surgery and manuscript revision.

ANZ: Radiologist reporting and manuscript revision.

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