Sphenoid Sinus Mucocele with Bilateral Blindness

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Abstract

Objective: We present a rare case of bilateral blindness in a young man with sphenoid sinus mucocele.

Case Summary: A male patient aged 18 years presented with three weeks history of worsening headache and vomiting. There was history of blurring of vision resulting in blindness in both eyes. He did not give any history of nasal or paranasal infection or fever. On general physical examination, there was no abnormality detected. Neurological examination revealed left ptosis, bilateral dilated pupils with afferent pupillary defect and no vision in both eyes. There was evidence of left complete ophthalmoplegia. On fundoscopy, there was bilateral papilloedema. Cranial CT scan showed a homogenously isodense well delineated lesion in the sphenoid area with lateral extensions. Also, cranial MRI revealed a non-enhancing sphenoid region lesion extending to sellar, suprasellar and parasellar areas. A diagnosis of sphenoid sinus mucocele with secondary infection was suspected. He underwent a navigation-guided transnasal sphenoidotomy. On the first post operative day, there was improvement in his visual acuity and afferent pupillary response returned on the left eye. Histology is in keeping with mucocele. He was discharged on the 8th post operative day on oral antibiotics and steroids for four weeks.

Result: His visual recovery has been rather slow probably due to prolonged compression of the optic nerves and optic chiasm.

Conclusion: Sphenoid sinus mucocele is one of the rare causes of blindness. Early diagnosis and prompt surgical treatment improve the outcome.

Keywords: Sphenoid sinus; Mucocele; Bilateral blindness; Transnasal sphenoidotomy

Introduction

Sphenoid sinus mucocele is a rare condition [1,2]. Blindness is one of the complications described [3–6]. It could be unilateral or bilateral. Late presentation is a major challenge as it leads to a poor outcome in the recovery of vision.

Case Report

A male patient aged 18 years presented with three weeks history of worsening headache and vomiting. There was history of blurring of vision resulting in blindness in both eyes. He did not give any history of nasal or paranasal infection or fever.

On general physical examination, there was no abnormality detected. Neurological examination revealed left ptosis (Figure 1), bilateral dilated pupils with afferent pupillary defect and no vision in both eyes. There was evidence of left complete ophthalmoplegia (Figure 2). On fundoscopy, there was bilateral papilloedema.

His cranial CT scan showed a homogenously isodense, well delineated lesion in the sphenoid region with lateral extensions (Figure 3). Also, cranial MRI revealed a thin walled grossly expanded sphenoid sinus harbouring a non-enhancing hyperintense lesion on both the T1 and T2 weighted sequences (Figures 4 and 5).

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and minimal restricted diffusion. It elevated and compressed the optic chiasm and the optic nerves (Figure 6).

His haemoglobin was 15 g/dl and total white blood cell count was 10,980 cells/mm³ and C-reactive protein was 20.8 mg/dl. A diagnosis of sphenoid sinus mucocele with secondary infection was suspected. He was commenced on intravenous ceftriaxone 2gm 12 hourly, and vancomycin 1gm 12 hourly, dexamethasone 4mg 6 hourly and analgesics.

He underwent a neuronavigation-guided transnasal sphenoidotomy and microscope-assisted drainage of the mucocele. At surgery, light yellowish mucoid fluid was drained. On the first post operative day, there was perception of hand movement in the left temporal visual field and afferent pupillary response recovered. Post-operative cranial MRI showed minimal residual collection in the sphenoid sinus (Figure 7).

Histology of tissue specimen revealed polypoid tissue lined by respiratory epithelium with mucosal gland proliferation in a mucoid oedematous and congested stroma. Microscopy, culture and sensitivity of intra-operative mucoid fluid yielded no bacterial growth. An acid fast bacillus was not detected. Visual evoked potential on the 7th post operative day showed bilateral retino-optic pathway disorder. He was discharged on the 8th post operative day on oral antibiotics and steroids for four weeks.

Result

His visual recovery was slow probably due to compression of the optic chiasm and the optic nerves. The bilateral retino-optic pathway disorder on visual evoked potential evaluation is of poor prognostic significance.

Discussion

Blindness in sphenoid sinus mucocele could be sudden or insidious with antecedent headache [4,7]. Sphenoid sinus mucocele could present with nasal complaint [5,8]. However, absence of nasal complaint does not preclude it [9] as in our index patient.

The clinical manifestations are essentially a reflection of the degree and duration of compression or stretching of soft tissue structures surrounding the sphenoid sinus. Therefore, it could present with diplopia, proptosis, Horner’s syndrome, multiple cranial nerve palsies and pituitary dysfunction [4].

Cranial neuropathies are features in as many as 50% of cases [10]. The cranial nerves involved are II through VI [1]. Oculomotor, abducens, and optic nerves are the commonly involved nerves [11,12]. Involvement of the oculomotor, trochlear and abducens nerves leads to ophthalmoplegia. Involvement of the optic nerve leads to blindness as in our index patient. Facial numbness could be
a presenting feature in involvement of first and second branches of the trigeminal nerve.

In our patient, there was subclinical secondary bacterial infection as evidenced by the elevated levels of C-reactive proteins and white blood cell count. However, microscopy and culture of intraoperative mucoid effluent yielded no bacterial organism probably due to sterilization by the administered parenteral broad spectrum antibiotics.

Brain CT scan is the imaging modality of choice for defining the extent of the lesion [13]. It also assists in surgical planning. There are three criteria for CT diagnosis of a mucocele: homogenous isodense mass, clearly defined margin and patchy osteolysis around the mass [14].

Brain MRI, on the other hand, can equally delineate the lesion in relation to the surrounding soft tissues. The MRI findings in mucocele are variable. They depend on water and protein contents. On T1-weighted MRI, high water content shows hypointensity and high protein content shows hyperintensity. On the other hand, on T2-weighted MRI, high water content shows hyperintensity and high protein content shows hypointensity. Two basic MR appearances were encountered: either moderate to marked signal hypointensity in the expanded sinuses on T1- and T2-weighted images or moderate to marked signal hyperintensity on T1- and T2- images [15]. MR contrast enhancement, if present, is usually peripheral. Low signal could be seen in both T1 and T2 in superimposed fungal infections [16].

Typically, most mucoceles tend to be fairly bright on T1-weighted images compared to the brain and iso-hypointense on T2-weighted images. It is a pathognomonic MRI finding for mucocele [14]. Insipidated proteinaceous content could be signal void on MRI but hyperdense on CT scan. Therefore, CT scan and MRI are complementary in complicated cases [17].

Transnasal approach is the surgical approach of choice [2,5]. It could be navigation guided, endoscopic or microscopic. The use of Caldwell-Luc approach through the maxillary sinus has been described [18]. Prognosis for recovery of vision is poor if the onset of visual loss is sudden or if the visual acuity is no perception of light preoperatively [19].

Conclusion

Blindness caused by sphenoid sinus mucocele is rare. It is preventable and redeemable. High index of clinical suspicion, early diagnosis, and prompt surgical treatment can improve outcome.

References


