A sphenoid sinus mucocele simulating as retro bulbar optic neuritis

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A 25-year-old male presented with complaints of sudden diminution of vision with pain on eye movement in the left eye which was diagnosed clinically as retro bulbar optic neuritis. However, magnetic resonance imaging (MRI) showed lesion consistent with sphenoid sinus mucocele. Early surgical removal of mucocele led to complete recovery of vision, contrast and visual field. A high index of suspicion is necessary for intracranial lesions in all cases of retro bulbar neuritis, especially those with atypical symptoms.

Key words: Magnetic resonance imaging, optic neuritis, sphenoid sinus mucocele, visual evoked response

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Optic neuritis (ON), an acute inflammatory disorder of the optic nerve, typically presents with sudden monocular loss of vision and eye pain in young adults. There are various causes which can lead to ON. The cause of ON in mucosal diseases is due to the course of the optic canal through it. It leads to slowly progressive painless loss of vision, which may terminate in optic atrophy.

Case Report

A 25-year-old man presented to us with complains of sudden diminution of vision in the left eye for four days associated with frontal headache and pain on eye movement with no signs of upper respiratory infection or sinusitis.

On examination he had a visual acuity of 20/60 in the left eye and 20/20 in right eye with a left relative afferent pupillary defect (RAPD) of 0.9 log unit. Eye movements were full in both the eyes. Fundus examination revealed normal disc in either eye [Fig. 1a and b]. Contrast sensitivity by Pelli Robson was 1.65 in the right eye and 0.90 in the left eye. Color vision was normal in both the eyes (Pseudo-isochromatic Ishihara plate). Visual evoked response (VER) showed amplitude of 10 micro volts and latency of 103 milliseconds in the right eye and 4 micro volts and 130 milliseconds in the left eye. Goldmann visual field charting was normal in the right eye with baring of blind spot in the left eye [Fig. 2a and b]. No other systemic or neurological deficit was present.

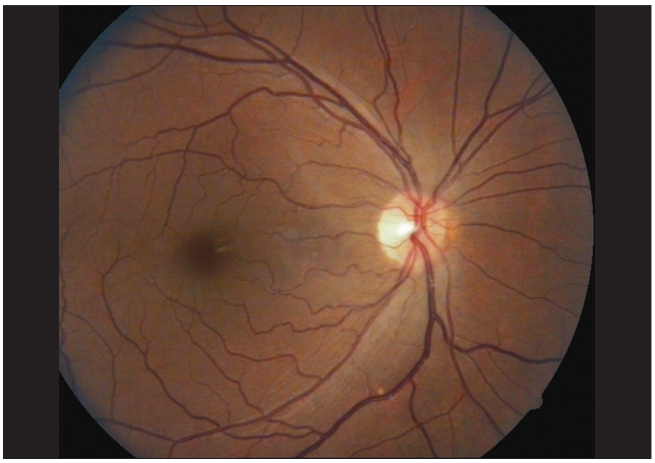

Figure 1a: Fundus photo of right eye showing normal optic disc

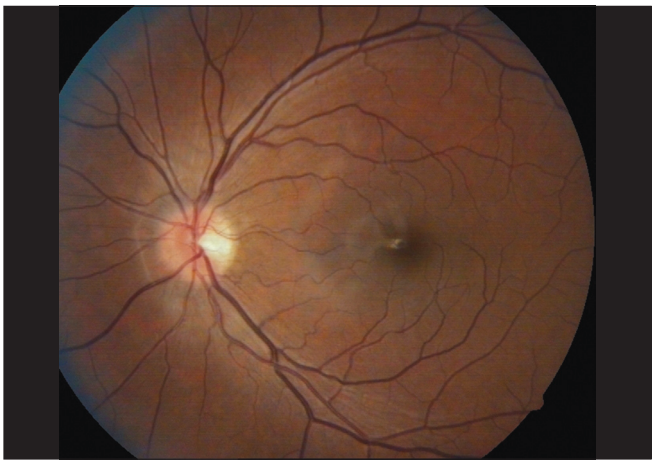

Figure 1b: Fundus photo of left eye showing normal optic disc and macula

A clinical diagnosis of retro bulbar optic neuritis was made on the basis of acute vision loss, pain on eye movement, affected contrast sensitivity and increased latency on VER.

The patient was asked to follow up after one week, at that time he complained of increase in ocular pain even at rest and his vision had dropped to 20/200. So to exclude any demyelinating disease magnetic resonance imaging (MRI) of the brain was advised. The MRI showed a large lobulated lesion, 41 × 66 × 64 mm along the floor of the sella displacing the pituitary superiorly with a suprasellar extension (no post-contrast enhancement). A diagnosis of sphenoid sinus mucocele was made [Fig. 3a and b]. The mass was large enough to be causing indentation of the cavernous sinus. The entire hormonal profile was done and all hormone levels in the blood were normal, including the serum prolactin level.

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The patient underwent a sub labial transseptal (SLTS) approach for excision of the sphenoid sinus mucocele under general anesthesia within two weeks of diagnosis. At three weeks postoperatively he had recovered normal visual acuity (20/20) but there was residual 0.3 log unit RAPD. The fundus examination was normal with no signs of optic disc pallor. The postoperative MRI done after three weeks showed no residual mucocele [Fig. 4a and b].

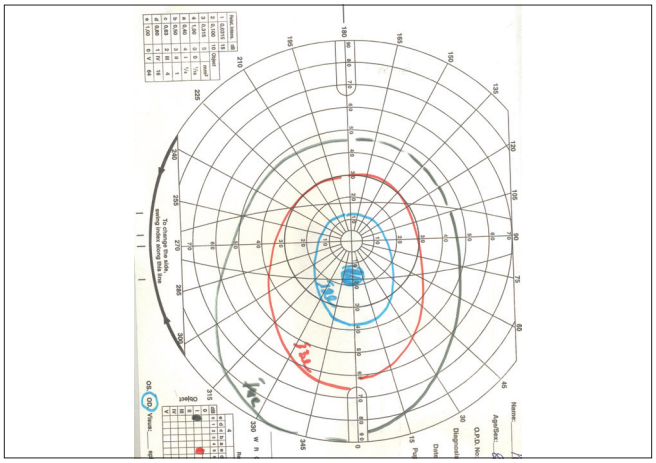

Figure 2a: Goldmann visual field charting showing normal visual field and blind spot in the right eye

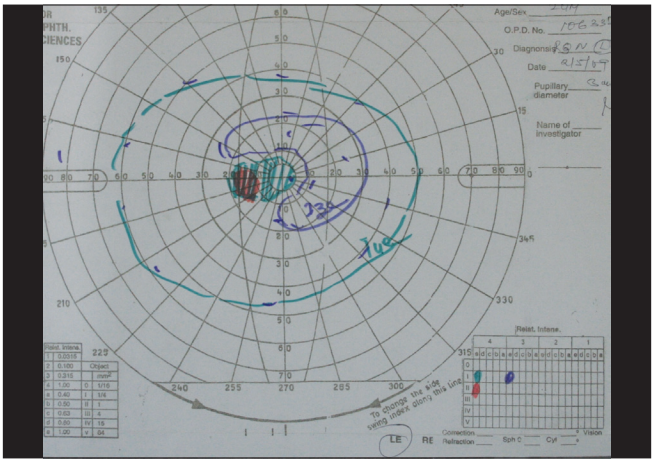

Figure 2b: Goldmann visual field charting showing (baring) and enlargement of blind spot size in the left eye

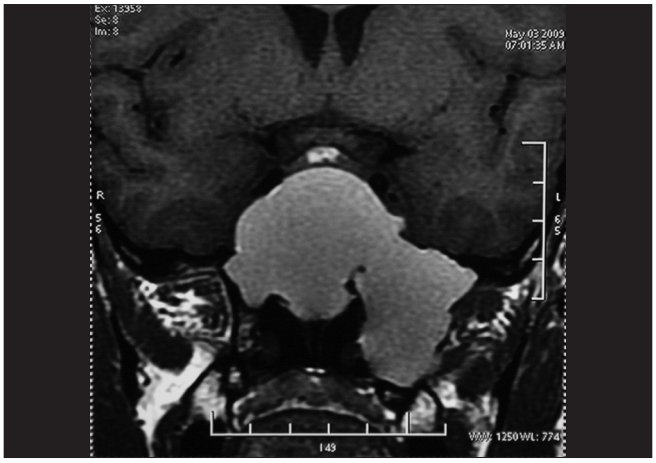

Figure 3a: Coronal image of patient showing T2 (hyperintense) lesion in the sphenoid sinus causing mass effect on the pituitary gland and the suprasellar cistern

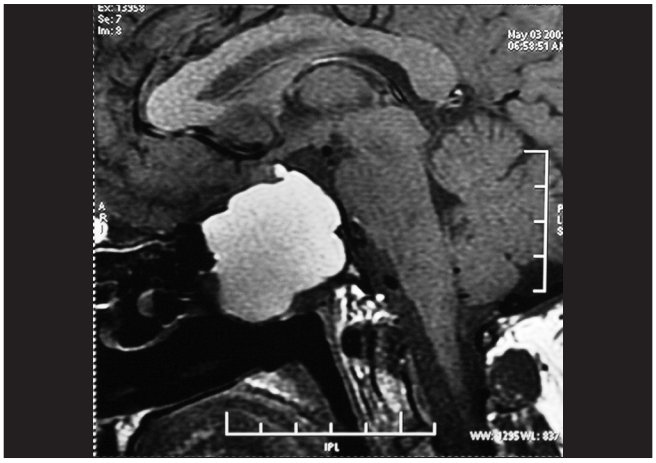

Figure 3b: Sagittal image showing T1 hyperintense lesion filling and expanding the sphenoid sinus

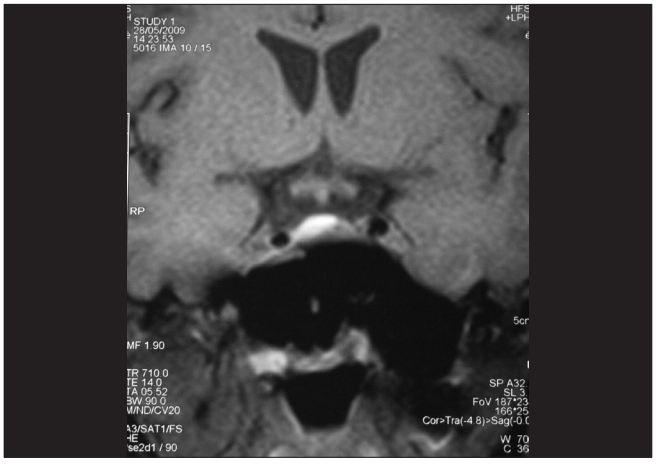

Figure 4a: Postoperative coronal image of the same patient showing absence of a mass lesion in the sphenoid sinus

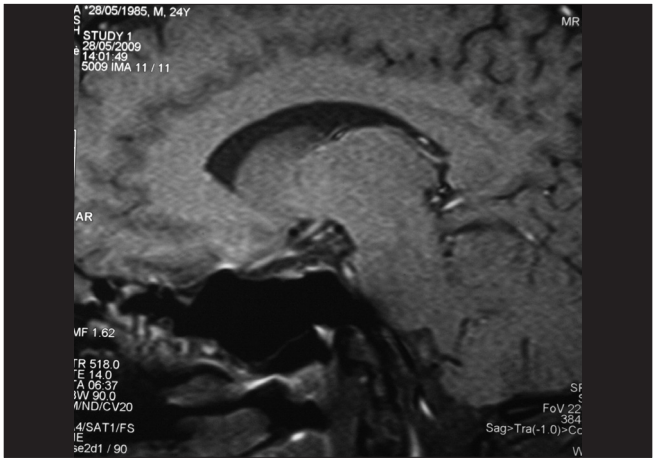

Figure 4b: Postoperative (sagittal) image of same patient showing normal aerated sphenoid with no residual lesion

Discussion

Optic neuritis, an acute inflammatory disorder of the optic nerve, typically presents with sudden monocular loss of vision and eye pain in young adults, more commonly in women and is usually associated with multiple sclerosis. Inflammation of the optic nerve can be due to infection; such as certain viral infections (mononucleosis), herpes zoster, and others, as well as granulomatous processes including sarcoidosis, syphilis, tuberculosis, Cryptococcus, and Lyme's disease and secondarily to contiguous spread of inflammation from the meninges, sinuses or orbit. However, often, retro bulbar neuritis could be mimicked by compressive lesions of the brain, adenexa and sinus diseases.[1]

The optic nerve is affected in mucosal diseases due to the course of the optic canal which is variable from person to person; in 51% it is in the lateral and superior wall of the sphenoid sinus, while in 25% cases bone dehiscence of the optic canal is found which leads to direct exposure of the nerve through a thin sinus mucosa.[2] Sphenoid sinus mucoceles are relatively rare, representing 1% of all paranasal sinus mucoceles.[3,4] They usually start unilaterally, but by the time of presentation, the entire sphenoid sinus complex may be opacified and expanded with thinning of its bony walls. Cranial neuropathies are a feature in as many as 50% of cases.[5]

In sphenoid mucocele the vision loss may be either due to the mass effect of the expanding mucocele leading to disturbance in the blood supply of the optic nerve or by the involvement of the optic canal resulting in unilateral, slowly progressive loss of vision, which may terminate in optic atrophy.[6,7] In our case the patient presented with sudden painful acute diminution of vision which may be explained by inflammation as well as the vascular compromised state of the optic nerve by a large mucocele. Treatment consists of marsupialization or surgical removal of the mucocele, usually resulting in rapid regression of the ophthalmic manifestations.[8] Optic neuropathy seldom recovers if there is a delay in surgery for more than 10 days.[6] However, with prompt surgical drainage complete visual recovery occurs as in our case.

In a resource-constrained country like India where neuroimaging is usually not done in cases of ON as the diagnosis is essentially clinical, it is judicious to advise neuroimaging in patients of retro bulbar ON. As in our case while the Goldman fields in the left eye were not conclusive, neuroimaging revealed the presence of a large mucocele with no sign of sinusitis, which was removed within a fortnight of diagnosis. This early diagnosis and prompt intervention helped in full visual recovery of the patient within a short period of one week.

Conclusion

A neuroimaging study should be considered essential in cases of retro bulbar neuritis even in the presence of a single atypical feature.

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