

Clinical Reasoning: An unusual pattern of optic disc swelling and visual loss

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SECTION 1

A 9-year-old boy was seen for bilaterally decreased vision. The patient noted gradual decrease in his visual acuity over a few months with difficulty seeing the blackboard. Otherwise, he was asymptomatic with no headaches. His examination showed visual acuities of 20/60 in his right eye and 20/400 in his left eye. For color vision, pseudo-isochromatic plates were 15/16 right eye and 0/16 in the left eye. There was a left relative afferent pupil defect. Ocular

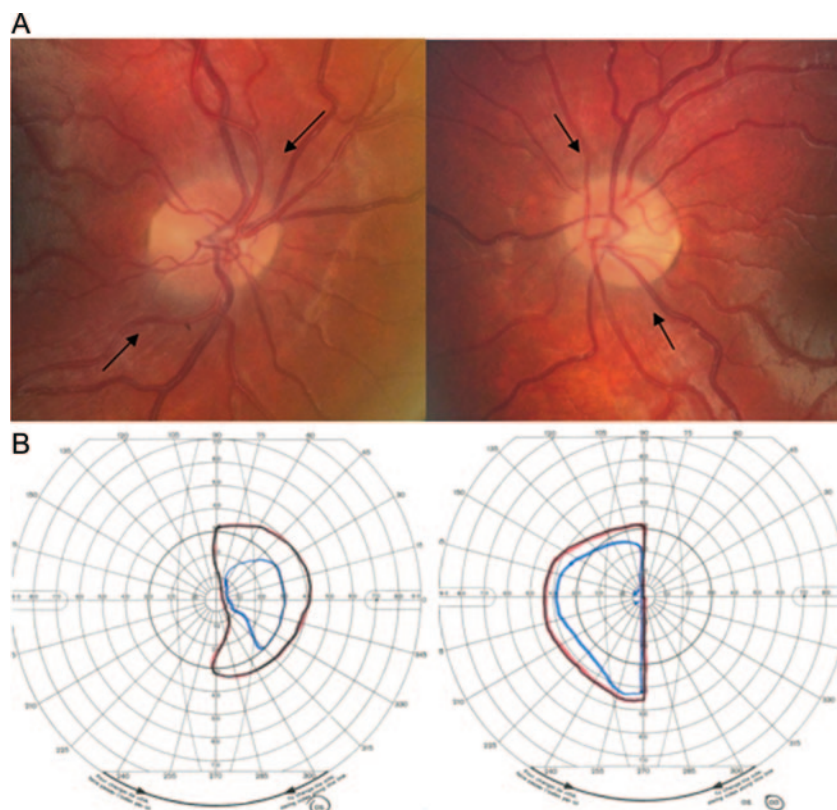
motility was normal. Funduscopy showed swelling of both optic discs affecting mainly the upper and lower aspects with sparing of the horizontal part of the disc, suggesting band atrophy. Maculae were normal (figure 1).

Questions for consideration:

1. What is the differential diagnosis?
2. What initial investigations would help you in narrowing the differential?

GO TO SECTION 2

Figure 1 Fundus photographs and Goldmann visual field



(A) Bilateral optic disc swelling affecting the upper and lower aspect of the optic discs (arrows). There is a horizontal band of atrophy that shows no swelling. Note the blurry margins of the upper and lower pole in contrast with the clear margins on the nasal and temporal aspects of the discs. (B) Goldmann visual field showing complete bitemporal hemianopia, with some involvement of the nasal field and macular region in the left eye, in keeping with the reduced acuity.

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SECTION 2

Optic disc swelling with visual loss is always a worrisome presentation. Optic neuritis in children is usually bilateral and associated with optic disc swelling more frequently than in adults.¹ The course is usually acute with ocular pain, instead of the painless, insidious presentation in this patient. Neuroretinitis, which is the association of optic disc swelling and a macular star (a ring of hard exudates in the macula), is usually due to infectious diseases such as cat-scratch disease, Lyme disease, syphilis, or toxoplasmosis.² Ischemia to the anterior optic nerve results in disc swelling and typically presents with segmental swelling affecting either the upper or lower half of the disc.³ However, outside of the setting of severe blood loss or hypotension, ischemic optic neuropathy is rare in children. Arterial hypertension can also present with bilateral disc swelling and also visual loss if there is secondary macular edema. However, retinal hemorrhages or exudates are usually present.⁴ Toxic, metabolic, nutritional, and hereditary optic neuropathies tend to present with central or cecocentral scotomas since they tend to selectively affect the papillomacular bundle,⁵ but not disc edema with the

exception of acute methanol toxicity. Infiltration of the anterior optic disc can cause swollen optic discs and visual loss. It usually occurs from leukemia, lymphoma, or conditions such as sarcoidosis. Compression of the optic nerve can also cause disc swelling. In our case with bilateral involvement, bilateral nerve compression or chiasmal involvement would be necessary to account for this presentation. Finally, papilledema from increased intracranial pressure should always be kept in mind in patients with disc swelling, although it does not commonly cause early acuity loss.⁶

In any patient with disc edema and visual loss, formal perimetry can be very useful in localizing the problem and narrowing the differential. In this case, Goldmann perimetry showed an almost complete bitemporal hemianopia. In addition to perimetry, patients with bilateral optic disc swelling require neuroimaging and lumbar puncture for measures of opening pressure and CSF content to address the above conditions.

Questions for consideration:

1. What would you do next?
2. What do you expect to find?

GO TO SECTION 3

SECTION 3

Asymmetric acuity and color vision loss, the presence of relative afferent pupillary defect, and bitemporal hemianopia all suggest asymmetric compression of the anterior visual pathway, namely the intracranial optic nerves and optic chiasm. An urgent head MRI showed a large suprasellar tumor extending into the third ventricle and causing hydrocephalus (figure 2).

Compression of the visual pathway requires urgent intervention to avoid permanent visual loss. The patient underwent urgent neurosurgery. The tumor

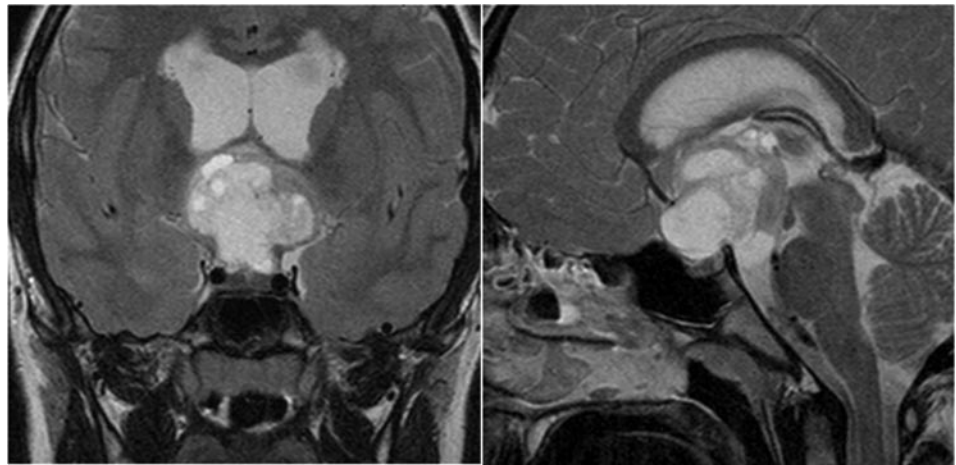
arose from the optic chiasm itself and pathology confirmed a pilocytic astrocytoma. Complete resection was not possible because of the risk of further damage to the visual pathway and therefore, the child was started on chemotherapy. A right occipital ventriculoperitoneal shunt was inserted.

Questions for consideration:

1. How would you explain the pattern of optic disc swelling?
2. What is the prerequisite for this to occur?

GO TO SECTION 4

Figure 2 T2-weighted MRI (left coronal image, right sagittal image) showing the suprasellar mass extending into the third ventricle and causing hydrocephalus



SECTION 4

This patient presented with hydrocephalus and bilateral papilledema. Increased intracranial pressure is transmitted to the optic nerve through the subarachnoid space and results in impaired axonal transport at the lamina cribrosa.⁷ Even though it is common for papilledema to begin in the upper or lower aspect of the disc, in this case the presence of pallor along the horizontal portion suggests previous axonal loss from the chiasmal tumor (i.e., primary atrophy). Damage to the retinal ganglion cells axons anywhere along their course results in retrograde degeneration that becomes evident on funduscopy 4 to 6 weeks after injury. The horizontal band or bowtie atrophy is due to the compromise of the nasal fibers that decussate in the optic chiasm.⁸ Thus papilledema is expressed mainly in the upper and lower aspects of the disc, which contain the arcuate fibers originating from the temporal aspect of the fundus.

DISCUSSION In this patient, there is a rare combination of signs. The presence of bilateral optic atrophy suggests chronicity since atrophy takes at least several weeks to develop. The bilateral bowtie pattern of atrophy localizes the lesion to the optic chiasm. As the tumor continued growing and extending into the third ventricle, it resulted in increased intracranial pressure, hydrocephalus, and papilledema. Papilledema is confined to the region containing axons that are not destined to decussate in the chiasm, because atrophied axons cannot swell. This particular type of swelling is known as twin peaks papilledema as it resembles 2 hills (the swollen peaks) separated by a valley (band atrophy).^{9,10} If papilledema persists it may also result in further axonal loss extending to the previously swollen aspects of the nerve, a secondary atrophy that will appear more diffuse without the localizing value of band or bowtie atrophy.

Additional asymmetric compression of the optic nerves explains the acuity loss, reduced color vision, and the left relative afferent pupil defect.

Detailed history and careful ophthalmoscopic examination provide valuable information about the disease course, the possible location of the lesion, and the underlying mechanism. This case is also a remainder that optic disc swelling should not be expected in the setting of optic atrophy. This concept is particularly important in patients with chronic papilledema and diffuse axonal loss: even if the intracranial pressure is significantly increased, there may be no changes in the appearance of the optic disc.

DISCLOSURE

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