Unilateral high myopia leading to asymmetric disc edema in idiopathic intracranial hypertension

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Asymmetrical disc edema in idiopathic intracranial hypertension (IIH) is an uncommon finding, with an unclear understanding. This report reinforces the importance of recognizing this entity in IIH diagnosis, and not misdiagnose the condition as unilateral disc edema. In this unique report, the causative association of asymmetric optic disc edema, with optic nerve kink, due to unilateral high myopia, is documented.

Key words: Asymmetric disc edema, high Myopia, idiopathic intracranial hypertension

Although uncommon, unilateral or asymmetric optic disc edema (difference of 2 or more grade between the two eyes) in idiopathic intracranial hypertension (IIH) is reported in the literature.[1-3] However, the exact mechanism for this phenomenon remains elusive. Herein, we report a case of IIH with marked asymmetric optic disc edema, with its unique association with an anatomic variation of optic nerve course in the orbit, due to unilateral high myopia.

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Case Report

A 25 years old lady (obese built) presented with esodeviation of her right eye for the past 1 month (December 2015). She had a faint horizontal doubling of images. In the past, she had refractive surgery procedure done in her right eye, elsewhere, 6 years back (no previous documents were available). There was no history of any specific antecedent medical illness or medication use. On examination, she had a best-corrected visual acuity (BCVA) of 6/60 in the right eye (−8 D/−1 D cyl/10°) and 6/6p in the left eye (Plano). The pupils were normally reacting to light and near stimuli and did not show any afferent defect. The color vision was normal (21/21) in the left eye, while in the right eye, the response was unreliable. Both eyes anterior segment exam (including intraocular pressure [IOP]-14 mmHg OD, 15 mmHg OS) were within normal limits. The orthoptic review revealed an esodeviation (25 PD for distance and near) with right eye minimal abduction deficit (−1 grade) [Fig. 1a-c]. On the fundus exam, the media were clear in both eyes. The right eye optic disc was hyperemic with trace disc edema (Frisen grading grade 0), while the left eye optic disc showed florid disc edema (Frisen grade 4), with peripapillary edema just stopping short of fovea [Fig. 1d and e], as described elsewhere.[1] The visual field assessment revealed no gross visual field defect

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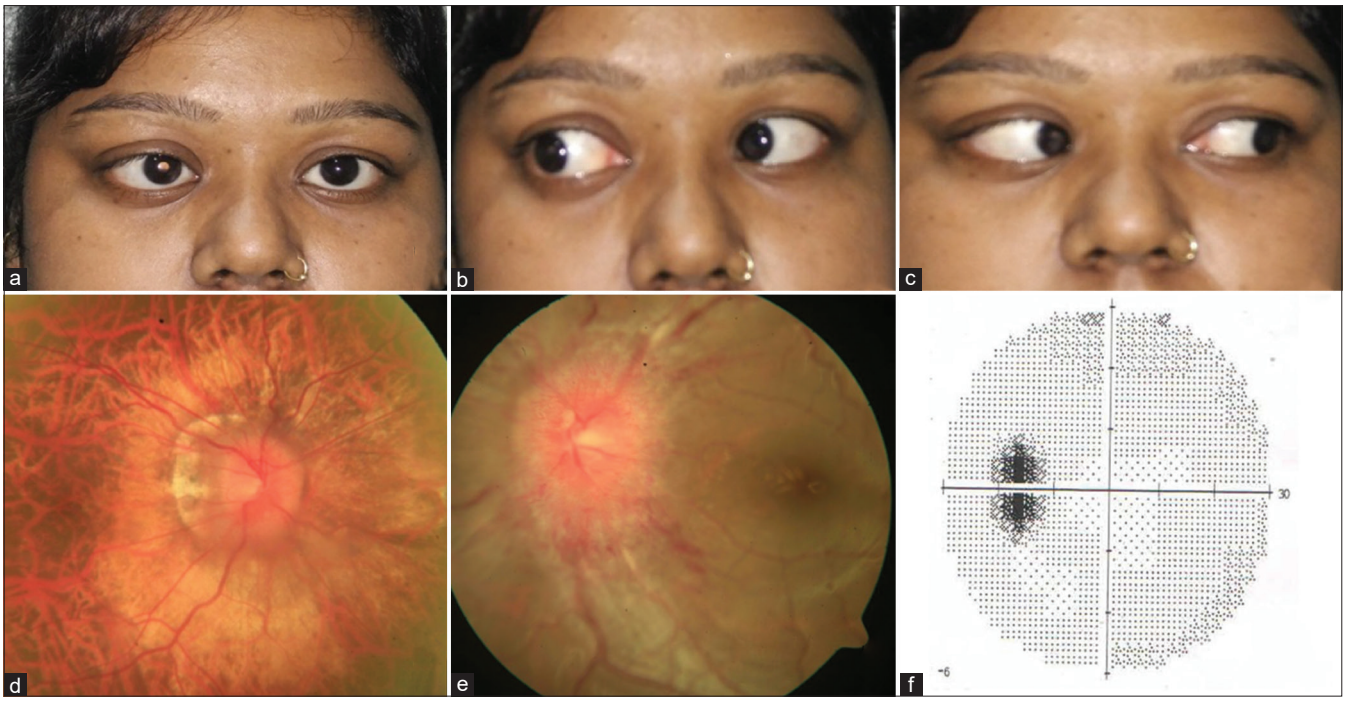

Figure 1: Clinical presentation: Esodeviation in primary gaze (a); minimal abduction deficit in dextroversion (b); normal abduction in levoversion (c) Asymmetric disc edema (d and e); Enlarged blind spot on automated perimetry of left eye (f)

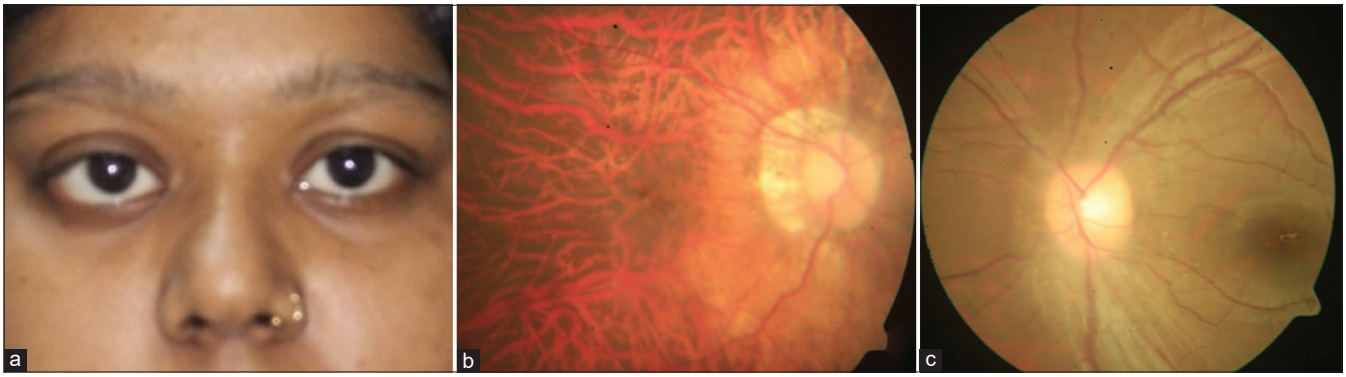

Figure 2: Outcome after medical management: Resolution of esodeviation (a) and disc edema, in both eyes (b and c)

in the right eye (confrontation field), while an enlarged blind spot was documented in the automated visual field of the left eye [Fig. 1f].

In a case of disc edema, the differential diagnosis and management varies, depending on whether the edema is unilateral or bilateral.[4] The marked asymmetry in disc edema in our case was perplexing. On a detailed exam, we noted the marked asymmetry in axial length (32.01 mm OD vs 23.18 mm OS). We proceeded on lines of papilledema management (considering the finding as asymmetric disc edema). Magnetic resonance imaging (MRI) of the brain ruled out any compressive pathology and subsequent, magnetic resonance venogram (MRV) ruled out any void in cortical venous sinuses. The patient was referred to the neurologist for cerebrospinal fluid (CSF) tap, which revealed an elevated opening pressure (280 mm of H_2O) and normal CSF analysis. A diagnosis of IIH was confirmed. The patient responded well to carbonic anhydrase inhibitor therapy (Tab acetazolamide 500 mg twice a day). The optic disc edema and esodeviation of the right eye resolved in about a month's time. [Fig. 2a-c] The resolved disc edema was maintained in both eyes at last follow-up (May 2019).

Discussion

As per current consensus, papilledema results in stasis of axoplasmic transport due to mechanical compression, leading to secondary vascular changes of venous dilation and hemorrhage.[5] Given this mechanism, the underlying reasons for asymmetrical papilledema remain unclear. In fact, in the IIH treatment trial, almost 7% of patients (n = 135) were having asymmetric optic disc edema.[1] Few hypotheses have been proposed to explain the atypical presentation of unilateral papilledema. Changes in the lamina cribrosa or optic nerve sheath anomaly were postulated as protecting the optic nerve from high intracranial pressure.[6,7] Tso *et al*. found that experimental elevated intracranial pressure can cause

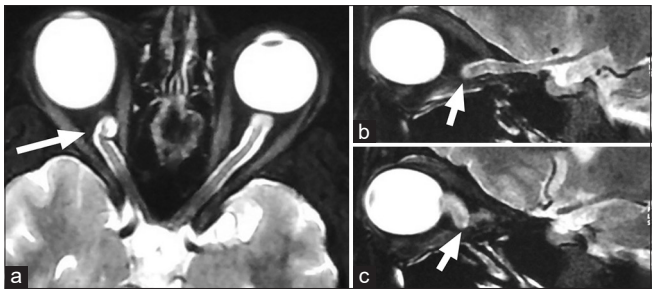

Figure 3: Kinking of right eye optic nerve (due to elongated globe of high myopia) causing mechanical obstruction of CSF flow (Arrows in a, b and c)

axoplasmic blockage at the level of the lamina cribrosa.[8] It is believed that the translaminar pressure gradient is modulated by the intraocular pressure anteriorly and the CSF-optic nerve subarachnoid pressure posteriorly.[9] However, as in our case, the asymmetric papilledema was not associated with differential intraocular pressure between the two eyes. Another hypothesis is a possible constriction of the bony optic canal on the side with less severe disc edema, but this could not be replicated by others.[2] CSF flow studies with computed tomography-assisted cisternography in IIH patients, who did not improve with medical therapy for intracranial pressure reduction, showed impaired contrast-loaded CSF flow into the subarachnoid space of optic nerve, indicating CSF compartmentation.[10] In our case, fine sections of orbital imaging revealed that a kink in the right eye optic nerve was present (presumably secondary to the enlarged right globe of high myopia) [Fig. 3a-c]. This kink in the right eye optic nerve was possibly mechanically obstructing the CSF flow to the optic nerve head (leading to only a trace optic nerve head swelling, in spite a raised CSF pressure). To the best of our knowledge, the kink in the optic nerve course, due to high myopia, leading to marked asymmetric optic disc edema in IIH, has not been documented yet in literature.

The learning pearl is that clinicians should not rule out the possibility of papilledema (bilateral disc edema), in cases of highly asymmetric swollen disks. Pseudo-disc edema can be ruled out by possibly adding fundus fluorescein angiography and or optical coherence tomography.[4] It is also useful to add contrast orbital imaging study, along with routine neuroimaging protocol, to view the course of the optic nerve, before labeling the disc edema as "unilateral."

Conclusion

Asymmetric disc edema is a possibility in IIH. The variation in the anatomic course of the optic nerve, especially in unilateral high myopes, could possibly cause mechanical obstruction of CSF and cause asymmetric optic disc edema.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Smith SV, Freidman DI. The idiopathic intracranial hypertension treatment trial: A review of the outcomes. Headache 2017;57:1303-10.
2. Bidot S, Bruce BB, Saindane AM, Newman NJ, Biousse V. Asymmetric papilledema in idiopathic intracranial hypertension. J Neuroophthalmol 2015;35:31-6.
3. Brosh K, Strassman I. Unilateral papilledema in pseudotumor cerebri. Semin Ophthalmol 2013;28:242-3.
4. Chiang J, Wong E, Whatham A, Hennessy M, Kalloniatis M, Zangerl B. The usefulness of multimodal imaging for differentiating pseudopapilloedema and true swelling of the optic nerve head: a review and case series. Clin Exp Optom 2015;98:12-24.
5. Hayreh SS. Optic disc edema in raised intracranial pressure. V. Pathogenesis. Arch Ophthalmol 1977;95:1553-65.
6. Lepore FE. Unilateral and highly asymmetric papilledema in pseudotumor cerebri. Neurology 1992;42:676-8.
7. Moster ML, Slavin M, Wall M. Unilateral disk edema in a young woman. Surv Ophthalmol 1995;39:409-16.
8. Tso MO, Hayreh SS. Optic disc edema in raised intracranial pressure. III. A pathologic study of experimental papilledema. Arch Ophthalmol 1977;95:1448-57.
9. Lawlor M, Zhang MG, Virgo J, Plant GT. Asymmetrical intraocular pressures and asymmetrical papilledema in pseudotumor cerebri syndrome. Neuroophthalmology 2016;40:292-6.
10. Killer HE, Subramanian PS. Compartmentalized cerebrospinal fluid. Int Ophth Clinics 2014;54:95-102.