CASE REPORT

Asymmetrical Intraocular Pressures and Asymmetrical Papilloedema in Pseudotumor Cerebri Syndrome

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ABSTRACT

This report is of two cases of asymmetrical papilloedema in patients with asymmetrical intraocular pressures (IOPs). The first patient presented with headaches, transient visual obscurations (TVOs), and elevated IOPs, and was found to have increased intracranial pressure caused by a torcula meningioma. He developed papilloedema after his IOPs were pharmacologically lowered; the papilloedema resolved after the IOP became elevated again after stopping his glaucoma drops, and then again returned as the IOP reduced when the drops were restarted. The second patient with a history of Sturge-Weber syndrome requiring previous left trabeculectomy, presented with left-sided TVOs, photopsia, and pulsatile tinnitus caused by idiopathic intracranial hypertension. Asymmetrical papilloedema was observed, worse in the eye with the lower IOP following trabeculectomy. These cases suggest that asymmetric IOP may be one factor that can influence the development of asymmetric papilloedema. Ophthalmologists finding disc swelling at low normal pressures should ask about symptoms of raised ICP, and neuro-ophthalmologists confronted with asymmetrical disc swelling should routinely measure IOP.

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Introduction

Pseudotumor cerebri syndrome most frequently presents with bilateral optic disc swelling; however, case reports and series have reported patients with either asymmetrical or truly unilateral swelling.^{1–6} The mechanism of this has not been fully elucidated, but one possibility includes a differential transmission of intracranial cerebral spinal fluid (CSF) pressure to the optic nerve head in an eye with relatively low intraocular pressure (IOP).⁷ It is postulated that the relative hypotony against an increased CSF pressure results in a net positive anterior translaminar pressure gradient at the level of lamina cribrosa, consequently inducing swelling at the optic disc.

Here we present two patients with lumbar puncture (LP)-confirmed elevated intracranial pressure (ICP) with markedly different IOPs and papilloedema between each eye.

Case reports

Case 1

A 46-year-old male nurse, with no known family history of ocular hypertension or glaucoma, presented to the eye emergency department with headaches and intermittent blurring of vision. On examination, his IOPs were 32 mm Hg in the right eye and 44 mm Hg in the left eye. His corrected acuity was 6/6 in each eye, with no relative afferent pupillary defect. His corneas were clear, and angles were open on gonioscopy. There was no pseudoexfoliation, inflammation, or any other anomaly of the anterior segment. Examination of the optic nerve heads showed no evidence of glaucoma and no evidence of optic disc swelling. Treatment was commenced with Travaprost 1 drop nocte to both eyes.

On review 1 month later, his IOPs were 22 mm Hg in the right eye and 28 mm Hg in the left eye. He was noted to have a swollen right optic nerve

Figure 1. Asymmetrical papilloedema developed one month after IOP lowering with travaprost eye drops.

head (**Figure 1**). He had full colour vision and visual fields in each eye. He was referred to the neuro-ophthalmology service where an additional finding of a soft tissue mass at the vertex of the patient's skull was noted.

Computed tomography (CT) identified an expansile midline soft tissue mass of the skull vault. It was noted to have intracranial and extracranial components and some bony spiculation. The mass was also displacing, extended into and partly occluding the superior sagittal sinus. There were other normal CSF spaces with normal appearances of the other venous sinuses. A lumbar puncture yielded an opening pressure of 42 cm CSF with normal constituents. The patient underwent a partial resection of the mass, and histology confirmed a World Health Organization (WHO) grade I meningioma. Treatment also included adjunctive intensity-modulated radiotherapy (50.4 Gy in 28 fractions).

Postoperative magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) demonstrated the partly excised dural-

based lesion extending into the superior sagittal sinus. Postero-inferiorly there was a lack of signal void within the sinus suggestive of occlusion or thrombosis; however, there were numerous parasagittal collateral venous structures seen bypassing the area. The anterior portion of the sinus was patent. There was no evidence of extension into the brain parenchyma and normal intracranial appearances elsewhere.

Given that the meningioma could not be completely excised, the patient was kept on acetazolamide 500 mg bd (twice daily) postoperatively as treatment for likely persistent disc swelling. One month postoperatively, he had bilateral asymmetrical disc swelling, worse in the right eye. IOPs were 15 mm Hg right and 20 mm Hg left. Visual fields were full. Three months after surgery, the swelling in his left eye had resolved, but the right eye remained swollen. His visual fields again remained full.

He continued on acetazolamide 500 mg bd, and ongoing reviews confirmed persistent low-grade swelling of the right optic nerve head and no swelling of the left optic nerve head (**Figure 2**).

Figure 2. Reduced papilloedema in the right eye on treatment with topical travaprost and oral acetazolamide.

Figure 3. Asymmetrical papilloedema in IIH with a previous left trabeculectomy.

On one review, his glaucoma medications were changed from Travaprost to timolol because of local side effects.

He attended a routine glaucoma follow-up 3 years post-surgery after forgetting to use the timolol eye drops while on holiday. He remained on acetazolamide 500 mg bd. IOPs were 25 mm Hg in the right eye and 32 mm Hg in the left eye. His visual field remained full, and on this examination he no longer had any swelling of either of the optic nerve heads. Timolol was recommenced once daily.

On treatment, his IOPs were 16 mm Hg in the right eye and 26 mm Hg in the left eye at 6-month follow-up. Again, his low-grade disc swelling returned, more pronounced in the right eye. Visual function remained normal with normal acuity, colour vision, and visual fields.

Case 2

A 30-year-old female was seen with transient visual obscurations (TVOs) in her left eye. Her medical history included Sturge-Weber syndrome with nevus flammeus affecting the left side of her face. At the age of 12, she had uncontrolled left ocular hypertension in association with her Sturge-Weber syndrome and underwent a left trabeculectomy. A review 9 years after trabeculectomy found corrected acuities of 6/6 on the right and 6/60 pinhole to 6/24 on the left. There was a full-thickness scleral hole and bleb leak, with IOP of 11 mm Hg in the right eye and 2 mm Hg in the left eye. She had hypotony maculopathy but no mention of disc swelling.

The patient underwent a bleb revision with a scleral patch graft and conjunctival closure. At 1 week, the IOP was 7 mm Hg on the left and at 6 weeks it was

10 mm Hg. The eye then remained stable on review, with good IOPs over the ensuing years.

She was seen again 9 years later complaining of left-sided photopsia and pulsatile tinnitus. She was in the first trimester of a pregnancy and had gained approximately 5 kg over the previous 2 months. She was not using any tetracyclines, vitamin A analogues, or any other relevant medications. She was not on any glaucoma medications, and IOPs were 19 mm Hg in the right eye and 10 mm Hg in the left eye. The trabeculectomy site looked healthy with no bleb leak. The left optic disc was swollen, and there was mild swelling of the right optic nerve head (Figure 3). Visual function remained normal: she had 6/6 acuity in each eye, normal colour vision with Ishihara plates, and normal visual fields with full threshold automated perimetry.

A non-contrast MRI, including time-of-flight sequences, did not show any abnormalities other than signs of elevated intracranial pressure: there was no evidence of venous sinus thrombosis. Bloods were unremarkable, with no evidence of anaemia. A lumbar puncture yielded an opening pressure of 35 cm CSF with normal CSF constituents. A diagnosis of idiopathic intracranial hypertension was made.

Discussion

A number of retrospective studies have identified a subgroup of patients with either unilateral or asymmetrical papilloedema. One report from a neurosurgical centre found that 4.4% of all papilloedema presentations were unilateral,⁸ and over a cumulative 24-year period in two tertiary neuro-ophthalmic practices, 15 cases of unilateral (not just asymmetrical) papilloedema were detected.⁵

The generally accepted mechanism of papilloedema is mechanical compression resulting in stasis of axoplasmic transport, leading to secondary vascular changes of venous dilation and haemorrhage.⁹ However, given this mechanism, the underlying reasons for asymmetrical papilloedema remains unclear.

Minckler et al. have shown in monkeys with light microscopy that optic disc oedema is histologically indistinguishable from swelling due to raised intracranial pressure.¹⁰ They also found that all eyes with lowered IOP developed disc swelling due to ocular hypotony, which resolved when the IOP returned to normal.¹⁰ These findings have been confirmed in humans, with a number of cases reporting the development of unilateral papilloedema from idiopathic intracranial hypertension (IIH) following surgical reduction of IOP for glaucoma.^{7,11,12}

Although neither of the larger case series looking at unilateral papilloedema reported the IOP of their patients,^{5,8} it is clear that asymmetrical IOP does not explain all cases of asymmetrical papilloedema. Bidot et al. found that the IOP differential between eyes in patients with asymmetrical papilloedema was not statistically significant.¹³ One possible explanation is the differential transmission of the elevated ICP to the optic nerve head. Huna-Baron et al. investigated whether the size of the intraorbital optic nerve sheath was different between eyes with asymmetrical disc swelling; however, no difference was found.⁵ More recently, Bidot et al. found that there was a wider bony optic canal in the eye of higher-grade papilloedema,¹³ raising the possibility that optic canal size may modulate the transmission of CSF pressure to the optic nerve head.

More recently, US National Aeronautics and Space Administration (NASA) astronauts on board the International Space Station for a mean of 6 months were shown to have an increased incidence of papilloedema.¹⁴ It is postulated that this may be contributed to by an altered translaminar pressure gradient in individuals subjected to zero gravity for a substantial duration.¹⁵ Under normal earth gravitational force, CSF pools in the lower spine, and CSF pressure at the eye level is on average 13 mm Hg lower than that location.^{16,17} In space, it is thought that the CSF is distributed evenly within the subarachnoid space compartments, which would increase the effective CSF pressure posterior to the lamina cribrosa, whereas their IOP remains unchanged.¹⁵

The existing case reports, and the two new patients we add to the literature, provide strong evidence that IOP can be an important factor in disc swelling. Both of our cases provide evidence that lowering IOP may “unmask” disc swelling from elevated ICP, by altering the translaminar pressure gradient: our second case corroborates this phenomenon that has previously been reported following trabeculectomy surgery.^{7,11,12} Our first case demonstrates that translaminar pressure is relevant to optic nerve head swelling when IOPs are above the reference range: the eye with normal IOP developed disc swelling, whereas elevated IOP in the contralateral eye appears to have prevented the optic disc from swelling. When his IOP increased after stopping topical antihypertensive medication, the disc oedema resolved. This was again reversed when he restarted his topical medications, and developed disc oedema once more.

Conclusion

A substantial difference in IOPs modulates the amount of swelling that occurs at each optic nerve head. The translaminar pressure gradient is modulated by the IOP anteriorly and the CSF optic nerve subarachnoid pressure posteriorly. Although not all cases of asymmetrical disc swelling in the context of raised intracranial pressure are caused by IOP differences, these cases help to provide an understanding of contributing factors to the underlying pathophysiological process.

From a clinical perspective, these findings suggest that ophthalmologists presented with disc swelling at low or normal pressures not usually associated with manifestations of hypotony should interrogate the patient for symptoms of raised ICP, and that neuro-ophthalmologists confronted with asymmetrical disc swelling should routinely measure the patients' IOPs. The observations also raise the possibility that one method of treating patients with papilloedema may be to marginally increase the IOP.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the article.

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