CASE REPORT

Intracanalicular Optic Nerve Swelling and Signal Change in Fulminant Untreated Idiopathic Intracranial Hypertension

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ABSTRACT

The role of the optic canal in the pathogenesis of papilloedema has been under scrutiny recently. Whether a larger canal precedes more severe papilloedema or is the result of bone remodelling from chronically raised pressure across a pressure gradient is not clear. The authors present the magnetic resonance imaging findings of a 29-year-old female with fulminant and untreated idiopathic intracranial hypertension. Imaging showed focal expansion and intrinsic signal changes of the intracanalicular optic nerve. The authors discuss the possibility of either fluid accumulation within the optic nerves from a water hammer effect across blocked optic canals resulting from the steep pressure gradient or opticomalacia (optic nerve softening) from chronic ischaemia.

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Introduction

Raised intracranial pressure is an essential feature of idiopathic intracranial hypertension (IIH). The role played by the optic canal in the transmission of this pressure to the optic nerve head has come under scrutiny recently.^{1–3} In a recent study, Bidot et al. have shown that a wide optic canal is associated with more severe papilloedema and visual loss, perhaps by less restriction to intracranial pressure transmission to the lamina cribrosa.¹ Whether the wider canal is secondary to bone remodelling from raised pressure or a congenital larger canal facilitates pressure transmission to the lamina cribrosa is unclear. Contrary to their findings, however, Sencer et al. have shown benefit in relieving papilloedema by endoscopic optic canal decompression in their cohort of 10 patients.⁴ This small, uncontrolled study has many limitations but does bring into question the role played by the optic canal in papilloedema severity and optic nerve functioning in raised intracranial pressure.

The physiologically narrow optic canal, by providing a communication between the larger basal cistern and the smaller peri-optic cerebrospinal fluid (CSF) space, is subject to the Venturi effect, which creates a

pressure gradient between these two spaces. According to the Venturi effect, when fluid flows from a larger-diameter tube to a smaller one, two changes occur: the pressure drops and velocity increases. Further elevation of the pressure in the larger-diameter tube causes further drop in the pressure within the narrower tube, thus steepening the pressure gradient even more. In IIH, owing to the much elevated intracranial pressure within the basal cistern, the pressure gradient is steepened between the basal cistern and the intracanalicular and orbital peri-optic spaces. We further postulate that in addition to the lowering of the intracanalicular pressure, there is a vacuum effect created by the lower pressure that pulls the surrounding optic nerve sheath inwards and contributes to “choking” of the optic nerve. We present the imaging findings that may be an example of this process in a patient with unabated IIH.

Case report

A 29-year-old female presented to Greys Hospital, a tertiary medical facility in KwaZulu-Natal, South

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Africa, with complaints of headache suggestive of raised intracranial pressure, tinnitus in the left ear, diplopia, and blurry vision. She had bilateral Frisén grade 4 papilloedema, a left sixth nerve palsy, and best-corrected visual acuity (BCVA) of 6/6 OU on Snellen chart. She had recent weight gain, the rate of which we were unable to quantify, but her body mass index (BMI) was 39.9 kg/m^2 (weight 92 kg, height 1.52 m). The computed tomography (CT) scan of her brain was normal, and venous sinuses were noted to be patent. The basic serum investigations, including iron studies, were normal. CSF pressure was 55 cm CSF in the left lateral position, CSF biochemistry was normal, and no cells were detected on microscopy. Routine testing of common central nervous system (CNS) infections was negative. A diagnosis of IIH was made based on the modified Dandy criteria, and she was commenced on acetazolamide, analgesics, and a weight loss dietary regime. Her symptoms improved after the lumbar puncture and commencement of medical treatment.

She was lost to follow-up for unclear reasons and presented 16 months later with complaints of worsening headaches and progressive left-sided visual loss of few months' duration. During this admission, she was noted to have a BCVA of 6/6 OD and 6/60 OS, bilateral pale and atrophic optic discs (Figure 1), and constricted visual fields (VF) OU to confrontational testing. Automated Humphrey VF was reliable OD (MD -23.03 DB , PSD 11.11 DB) and confirmed the field constriction (Figure 2). Automated Humphrey VF was

unreliable OS owing to her poor BCVA (MD -33.31 DB , PSD 2.42 DB). She still had the left sixth nerve palsy, and her BMI remained high at 40 kg/m^2 (weight 93 kg, height 1.52 m). The repeat CSF pressure measurement was 35 cm CSF, acellular on microscopy, and chemistry was normal. CSF fluorescent treponemal antibody absorption test, cryptococcal antigen test, GeneXpert test for tuberculosis, neurotropic virus tests (cytomegalovirus, herpes simplex virus 1 and 2, varicella-zoster virus), and cytology for malignancy were all negative. She was recommenced on acetazolamide and analgesics and noticed once again improvement of her symptoms following the lumbar puncture. The headaches and left esotropia improved, and BCVA improved to 6/36 OS on review 2 weeks later. Medical treatment was continued, and surgical intervention was not offered in view of her now atrophic optic nerves.

Her brain magnetic resonance imaging (MRI) and magnetic resonance venogram done following the second admission and prior to the lumbar puncture when she was still symptomatic were normal. Her orbit MRI showed bilateral optic atrophy but, in addition, focal expansion of the intracanalicular optic nerves affecting the left optic nerve more than the right. There was also asymmetrical T2 hyperintensity within the focal expansion demonstrated by acquiring 0.7-mm slices with a three-dimensional (3D) T2 turbo spin echo sequence on a 1.5-T MR scanner (Figures 3 and 4). In IIH, signal changes within the optic nerve are atypical, unlike optic

Figure 1. Bilateral secondary optic atrophy from chronic raised intracranial pressure.

Figure 2. 30-2 Humphrey visual field (HVF) of the right eye demonstrating constriction. HVF of the left eye was unreliable owing to poor acuity but was also constricted on confrontational testing. Right eye: MD -23.03 DB, PSD 11.11 DB; left eye: MD -33.31 DB, PSD 2.42 DB.

Figure 3. High-resolution magnetic resonance 3D T2-weighted 0.7 mm axial slices at the optic canal level showing focal optic nerve expansion and hyperintense signal changes within the optic nerves bilaterally (white arrows).

neuritis and ischaemic optic neuropathy where T2 changes may be evident during the acute stage from inflammation or oedema and during the chronic stage from gliosis or opticomalacia (softening of the optic nerve from compression-induced ischaemia). The signal change present within the intracanalicular optic nerves in this patient with IIH, even if subtle, was still abnormal. We propose that in this patient, the T2 signal change seen within the expanded optic nerves was qualitatively indicative of either accumulated CSF fluid or opticomalacia from compression-induced ischaemia.

Discussion

The term optic nerve compartmentalisation has been reported by various authors as representing either compartmentalisation between the intra-ocular pressure and peri-optic CSF pressure at the optic nerve head as in glaucoma⁵ or between the intracranial subarachnoid space (SAS) and the peri-optic SAS at the optic canal as in IIH and cryptococcal meningitis.^{6,7} In the following discussion, we refer to compartmentalisation at the optic canal as it occurs in cryptococcal meningitis and perhaps IIH too.

The Venturi effect across the optic canal in papilloedema creates a steep pressure gradient as long as CSF flow across the canal remains unobstructed. Compartmentalisation, however, suggests

Figure 4. High-resolution magnetic resonance 3D T2-weighted 0.7 mm coronal slices. (a) Coronal images of the intra-orbital optic nerve. (b) Hyperintense signal within the right intracanalicular optic nerve (solid arrow). (c) Hyperintense signal within the left intracanalicular optic nerve (fragmented arrow). (d) Coronal images of the intracranial optic nerves. The arrowheads in a and d point to normal optic nerve signal.

Figure 5. Hypothetical stages in the development of intracanalicular optic nerve swelling and signal change in untreated idiopathic intracranial hypertension (IIH). Reversibility is possible at stages one and two if the CSF pressure is lowered. The presence or absence of bone remodelling from chronically elevated CSF pressure accounts for asymmetrical papilloedema in IIH.

blockage at the optic canal and separation of the peri-optic CSF compartment from the intracranial basal cistern CSF compartment.

Killer et al. have proposed a plausible mechanism to explain asymmetrical or unilateral papilloedema in IIH. Varying optic nerve sheath

and restrictive subarachnoid elements comprising septae, trabeculae, and pillars possibly limit the pressure to the less affected optic disc.^{6,8,9} In addition, raised intracranial pressure induces meningotheelial cell proliferation in the optic canal, thus contributing to the optic canal “choking” of the optic nerve.² In a case report and review of *Cryptococcus*-induced visual loss, we too have been able to demonstrate compartmentalisation between the intracranial SAS and the peri-optic SAS caused by a tight blockage at the level of the optic canal that subsequently re-opened with lowering of intracranial pressure.^{7,10} A pressure valve mechanism seems to operate when the pressure gradient is very steep and is re-opened when the gradient is lowered by lowering of the intracranial pressure.

We postulate that during the height of our patient’s illness, a steep gradient of CSF pressure developed across the optic canal resulting in a pressure valve mechanism, similar to the cryptococcal meningitis scenario. Signal changes within the intracanalicular optic nerve developed from two possible mechanisms resulting in either CSF accumulation within the nerve or opticomalacia from chronic ischaemia. Episodes of Valsalva could possibly have caused the forced egress of CSF across this blockage and very similar to the mechanism proposed by Gardner¹¹ for syringomyelia in Chiari I malformation, causing accumulation of fluid within the optic nerve. The focal expansion of the intracanalicular optic nerve is supportive of a space-occupying fluid-filled cavity (syrinx) within the optic nerve. An alternative explanation for the intracanalicular optic nerve signal change is chronic ischaemia. Chronic compression or “choking” of the optic nerve caused by the steep pressure gradient and meningotheelial proliferation caused vascular compromise and optic nerve ischaemia. Chronic ischaemia, however, cannot explain the focal expansion of the nerve. Standard T2 and T2 FLAIR MRI with fat saturation and 3-mm slices were insensitive in detecting the signal change within the optic nerves owing to the small size of the signal and the surrounding bony artefact. However, by acquiring 0.7-mm slices with a 3D T2 turbo spin echo sequence on the 1.5-T MR scanner, we were able to demonstrate this focal expansion and hyperintensity within the intracanalicular optic nerve on axial and coronal images (Figures 3 and 4). Atrophy of the intra-orbital optic nerve

emphasises this finding. Signal intensity measurement of the intracanalicular hyperintensity was equivalent to that of the peri-optic CSF (759 and 743 units, respectively), suggesting a fluid-filled cavity within the optic nerve. Unfortunately, surrounding bony artefact precluded diffusion-weighted imaging of the intracanalicular optic nerve.

The discussion presented by Killer, Bidot, and Mader on the role played by the optic canal in papilloedema severity certainly provides much insight into the structural dynamics and CSF pressure across the optic canal in IIH.^{2,3,12} Bidot’s suggestion that larger-diameter optic canals are associated with more severe papilloedema and visual field loss and that smaller optic canals protect the optic nerve head from raised intracranial pressure is supported by Killer, who adds that even if the bony canal is not narrow, the compartmentalisation and protection might be provided by meningotheelial proliferation and thickening of the meninges within the canal secondary to raised intracranial pressure. The effect of chronic compartmentalisation is unknown. Mader, however, is not supportive of their theory. He asserts that asymmetrical papilloedema affecting the right eye more commonly and present even after return of CSF pressure to normal in astronauts cannot be accounted for by canal size alone.

We would like to add to that discussion by proposing three stages of pressure effect on the intracanalicular optic nerve in unabated IIH and any cause of chronically raised intracranial pressure (Figure 5). During the first stage, there is steepening of the pressure gradient across the optic canal. The initial small pressure gradient does not impede transcanalicular CSF flow, thus permitting transmission of the high intracranial pressure to the lamina cribrosa and papilloedema results. As the intracranial pressure increases, a critical pressure gradient is reached when a valve-like mechanism develops. The pressure valve is created by subarachnoid trabeculae, septae, pillars, and hyperplastic meningotheelial cells as described by Killer.^{2,9} At this second stage, there is limitation of intracranial pressure transmission to the lamina cribrosa produced by this intracanalicular pressure valve. However, re-opening of this valve is potentially possible by lowering intracranial pressure and decreasing the pressure gradient, as we have shown in the case of cryptococcal meningitis.⁷ With chronically raised pressure, a third stage occurs when

depending on the malleability of the peri-canalicular bone, bone remodelling from the raised pressure creates canal widening. Differential canal widening between the two sides perhaps accounts for asymmetrical papilloedema. If the peri-canalicular bone remains non-pliable, the canal remains narrow and the pressure effect is then directly transmitted onto the intracanalicular optic nerve. Chronic pressure or “choking” of the intracanalicular optic nerve can cause vascular compromise and sudden onset loss of central vision from posterior ischaemic optic neuropathy or opticomalacia from chronic ischaemia. Furthermore, tiny fluid-filled cavities within the intracanalicular optic nerve are likely during this stage when the optic canals remain narrow. They conceivably arise from chronically elevated intracranial CSF pressure extruding CSF into the optic nerve during Valsalva, similar to Gardner’s¹¹ explanation for a water hammer effect that causes syringomyelia in Chiari I malformation. Unlike syringomyelia, which impairs functioning of the crossing spinothalamic fibres, none such crossing fibres exist within the intracanalicular optic nerve and hence visual disturbance is less likely or minimal from this cavitation. Regression of the optic nerve fluid-filled cavity is also likely with lowering of intracranial pressure, as occurs with syringomyelia following posterior fossa decompression for Chiari I malformation.

The above-described stages are hypothetical but do offer some explanation for previous observations.^{1,7} Our case provides some evidence for blockage of CSF flow at the optic canal in raised intracranial pressure and gives credence to the compartmentalisation in IIH alluded to by Killer.² The focal expansion of the intracanalicular optic nerves with intrinsic signal changes on high-resolution MRI scan do point to the optic canals playing a meaningful role in optic nerve dysfunction in unabated raised intracranial pressure in IIH. This abnormality is perhaps missed on routine MRI scans either from too large slices selected, the timing of imaging, or collapse of these spaces post lowering of intracranial pressure. Focal expansion of the nerve is more supportive of fluid accumulation rather than chronic ischaemic changes. However, this possibility needs to be further investigated, and we suggest the use of the 3D T2-weighted turbo spin echo high-resolution sequence with 0.7-mm slices preferably on a 3-T MRI scanner.

Declaration of interest

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

References

- [1] Bidot S, Clough L, Saindane AM, Newman NJ, Biousse V, Bruce BB. The optic canal size is associated with the severity of papilledema and poor visual function in idiopathic intracranial hypertension. *J Neuro-ophthalmol* 2016;36:120–125.
- [2] Killer HE. Asymmetric papilledema in idiopathic intracranial hypertension: comment. *J Neuro-ophthalmol* 2015;35:330–331.
- [3] Mader TH, Gibson CR, Hart SF, Lee AG. Asymmetric papilledema in idiopathic intracranial hypertension: comment. *J Neuro-ophthalmol* 2016;36:111–112.
- [4] Sencer A, Akcakaya MO, Basaran B, Yorukoglu AG, Aydoseli A, Aras Y, F. Sencan, B. Satana, I. Aslan, O. F. Unal, N. Izgi, and A. Canbolat. Unilateral endoscopic optic nerve decompression for idiopathic intracranial hypertension: a series of 10 patients. *World Neurosurg* 2014;82:745–750.
- [5] Orgul S. Compartment syndrome in the optic nerve: a new hypothesis in the pathogenesis of glaucoma. *Acta Ophthalmol* 2012;90:686–689.
- [6] Killer HE, Jaggi GP, Flammer J, Miller NR, Huber AR, Mironov A. Cerebrospinal fluid dynamics between the intracranial and the subarachnoid space of the optic nerve. Is it always bidirectional? *Brain* 2007;130(Pt 2):514–520.
- [7] Moodley A, Naidoo N, Reitz D, Chetty N, Rae W. The optic nerve compartment syndrome in *Cryptococcus*-induced visual loss. *Neuro-Ophthalmology* 2013;37:124–128.
- [8] Killer HE, Jaggi GP, Miller NR, Huber AR, Landolt H, Mironov A, P. Meyer, and L. Remonda. Cerebrospinal fluid dynamics between the basal cisterns and the subarachnoid space of the optic nerve in patients with papilloedema. *Br J Ophthalmol* 2011;95:822–827.
- [9] Killer HE, Laeng HR, Flammer J, Groscurth P. Architecture of arachnoid trabeculae, pillars, and septa in the subarachnoid space of the human optic nerve: anatomy and clinical considerations. *Br J Ophthalmol* 2003;87:777–781.
- [10] Moodley A, Rae W, Bhigjee A. Visual loss in HIV-associated cryptococcal meningitis: a case series and review of the mechanisms involved. *S Afr J HIV Med* 2015;16:1–9.
- [11] Gardner WJ. Hydrodynamic mechanism of syringomyelia: its relationship to myelocoele. *J Neurol Neurosurg Psychiatry*. 1965 Jun. 28:247–59.
- [12] Bidot S, Bruce BB, Saindane AM, Newman NJ, Biousse V. Asymmetric papilledema in idiopathic intracranial hypertension: response. *J Neuro-ophthalmol* 2015;35:31–36.