

Case Report

Optic fenestration in benign intracranial hypertension: case report

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ABSTRACT

Condition of raised intracranial pressure without any mass lesion and normal cerebrospinal fluid composition is termed as idiopathic intracranial hypertension or pseudotumor cerebri. Raised intracranial tension with visual effects was treated by trans-sphenoidal optic fenestration as an emergency intervention to salvage the vision. The lamina papyracea on either side was lifted off the orbital periosteum and thick bone of the ethmoid sphenoidal junction was drilled with a diamond burr to thin it and elevate the bone covering the optic nerve.

Keywords: Benign intracranial hypertension, Visual impairment, Trans sphenoidal, Optic fenestration

INTRODUCTION

Heinrich Quincke in 1893 first reported idiopathic intracranial hypertension and termed it as meningitis serosa. The term "pseudotumor cerebri" was introduced in 1904, while benign intracranial hypertension in 1955.¹

The arachnoid membrane of the optic nerve is continuous with the arachnoid membrane of the subdural intracranial space and thus allows for the raised intracranial pressure to be transmitted to the subarachnoid space within the optic nerve sheath resulting in optic nerve head edema.

Nerve fiber dysfunction due to axonal swelling can result in loss of central vision, a decrease in peripheral vision, and, ultimately, optic atrophy.² Symptoms in benign intracranial hypertension (BIH) are non-specific and are those of increased intracranial pressure. Headaches, nausea/vomiting, and visual disturbances are the most common presenting symptoms. Headaches are predominantly frontal in location, becoming worse on lying down.³

The diagnosis of benign intracranial hypertension is based upon the modified Dandy criteria, which includes - signs and symptoms only consistent with increased intracranial pressure (ICP) (i.e. headaches, nausea, vomiting, transient visual obscurations, papilloedema), no localizing focal neurological signs except unilateral or bilateral sixth nerve paresis or other signs associated with increased ICP, cerebrospinal fluid (CSF) opening pressure ≥ 25 cm of water (20-25 cm of water are borderline measurements) and without CSF cytological or chemical abnormalities and normal neuroimaging adequate to exclude cerebral venous thrombosis (i.e. usually magnetic resonance imaging combined with magnetic resonance venogram).⁴ Imaging features that support the diagnosis of idiopathic intracranial hypertension include; prominent subarachnoid space around the optic nerves, papilloedema i.e. flattening of the posterior sclera or enhancement of the prelaminar (intraocular) optic nerves, enlarged arachnoid outpouchings, bilateral venous sinus stenosis.^{5,6}

Medical management includes the carbonic anhydrase inhibitors, loop diuretics and topiramate. Surgical intervention should be considered on failure of conservative medical treatment and when the onset of the disease is acute and severe with rapid progression.⁷

Temporary measures include the serial lumbar punctures or a continuous lumbar drain as the CSF reforms rapidly.

Long-term surgical treatment includes optic nerve sheath fenestrations (ONSFs) or CSF shunts; lumbo-peritoneal shunt (LPS), ventriculo-peritoneal shunt (VPS), or ventriculo-atrial shunt (VAS).

CASE REPORT

A 47 years old lady was admitted in the neurology emergency services of a tertiary care facility with bilateral severe supraorbital ache and blurring of vision. The blurring was progressively worsening on either side for the last 3 months. There was no relief with, carbonic acid anhydrase inhibitor acetazolamide that reduces CSF production. Magnetic resonance imaging showed features of raised intracranial tension, namely enlarged arachnoid outpouchings with absence of any mass lesion (Figure 1).



Figure 1: MRI image.

The ophthalmologist evaluation's showed bilateral retinal fibre layer thinning (optic nerve degeneration) with grade 4 papilloedema on fundus examination with tunnel vision. Extraocular movements were normal. Rest of the neurological examination was within normal limits. Perimetry showed central vision sparing with loss of peripheral vision. Patient was referred for endoscopic trans naso-ethmoido-sphenoidal fenestration of the optic nerve sheath. Under general anaesthesia an endoscopic middle meatus antrostomy and fronto ethmoidectomy was performed. A posterior septectomy and wide sphenoidotomy was completed. The lamina papyracea on either side was lifted off the orbital periosteum in the region of the posterior ethmoids. The elevation was extended posteriorly crossing the orbital apex to reach the sphenoidal roof. The thick bone of the ethmoid sphenoidal junction had to be drilled with a diamond burr to thin it and elevate the bone covering the optic nerve (Figure 2).

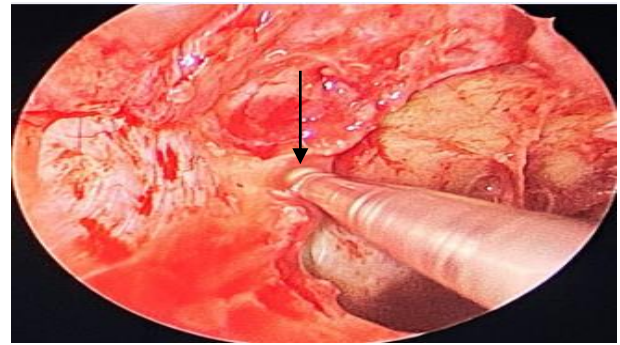


Figure 2: Drill over bone at ethmoido-sphenoidal junction over the optic nerve.

The entire orbital apex and the optic nerves on the sphenoidal roof were delineated (Figure 3). The dural sheath/meninges over the optic nerve was slit and fenestrated using a sharp no. 12 sickle shaped blade. (Figure 4) to facilitate a free flow of cerebrospinal fluid (Figure 5) inside the sphenoid.

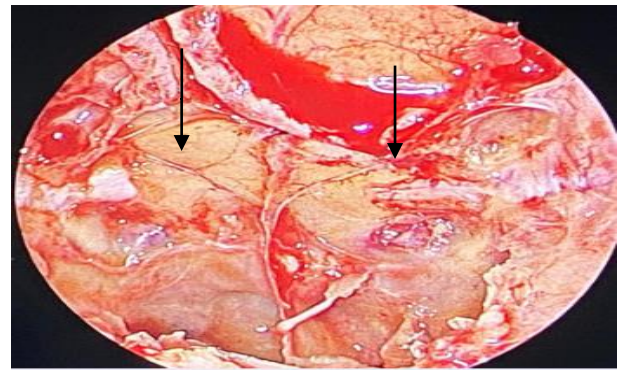


Figure 3: Bilateral exposed optic nerves after bone removal.

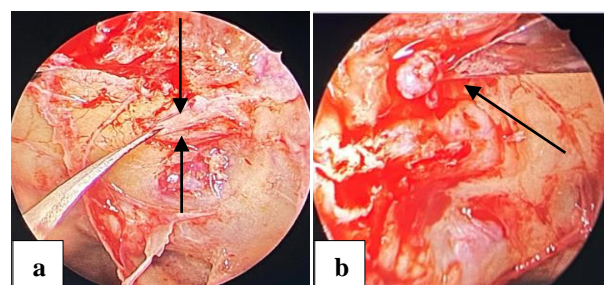


Figure 4: (a) sheath over left optic nerve being fenestrated, and (b) sheath over right optic nerve being fenestrated.

Pupils were reactive and patient shifted to the recovery. Nasal tamponade was obtained with inflatable airway merocels. The post-operative phase was uneventful with normal perception of light; the packs were removed on second day. Blurring and headache receded on day two of the surgery.

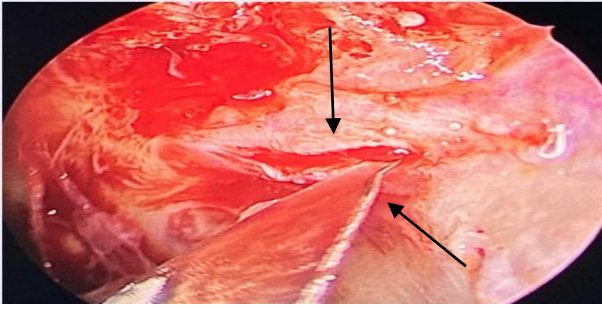


Figure 5: CSF egress after optic nerve sheath is cut.

DISCUSSION

De Wecker described the optic nerve fenestration procedure in 1872 as a technique to relieve optic nerve head (ONH) edema by incising the meninges surrounding the optic nerve.⁸

Once the bone around the optic nerve is drilled, the local CSF pressure decreases, but still, the subarachnoid space remains patent. As the sheath is opened as in optic nerve fenestration procedure, a local fistula is created for persistent CSF egress, so the pressure in optic nerve head further decreases. After 48-72 hours, fibroblasts proliferate, not only sealing the fistula but also filling the subarachnoid space permanently. Thus, the raised CSF pressure is not directly transmitted to the optic nerve fibres or the optic nerve head, and papilloedema resolves.^{9,10}

Before taking up the patient for optic nerve fenestration surgery patients must undergo a complete ophthalmic history, neurologic examination, and an ophthalmic examination including visual acuity, pupillary assessment, visual field testing, and fundoscopic examination to evaluate for presence and extent of papilledema (swelling of optic nerve). Emergent neuroimaging (CT scan or MRI) should be performed to rule out any intracranial processes such as dural sinus thrombosis, which can result in papilledema. Lumbar puncture must be done to check the CSF composition and opening pressure. Preoperative discussion should include reviewing the risks of orbital surgery in such a proximity to the optic nerve and the need for continued treatment of the underlying disease process, as this procedure is not a cure for the disease.

The optic nerve fenestration surgery can be performed via the trans-nasal endoscopic route and trans-orbital route. The three main transorbital approaches for ONSF are superior eyelid, lateral orbital and medial transconjunctival.¹¹

In a meta-analysis by Feldon et al based on retrospective data of ONSF performed via the transorbital route on 423 eyes in patients with IHH, visual acuity improved in 50% of eyes and visual field improved in 72% of eyes post surgery.¹²

The complication rate of transorbital optic nerve fenestration surgery was found to range broadly between 4.8-45% with a mean of 12.9% in the study by Lee et al.¹³ Complications reported with transorbital approaches which include; diplopia, which is typically temporary, sudden loss of vision (either from a vascular occlusive event, direct injury to nerve, or a hemorrhage), orbital or intra-sheath hemorrhage resulting in vision loss and pupillary abnormality from damage to para-sympathetic nerve fibers.¹¹ Scarring, fibrosis, recti muscle injury and fat adhesion are common sequelae of the above transorbital approaches.¹⁴

With trans-nasal endoscopic procedure there was better visualization of the neurovascular structures. The procedure is cosmetically appealing with shorter hospital stay of the patient and most importantly bilateral access may be gained through minimally invasive surgery.¹⁴

Endoscopic trans-nasal surgery deals with the local CSF pathophysiology alterations, offering a bilateral optic nerve sheath decompression utilizing the same corridor as in our case bilateral optic nerve fenestration was performed.

The only limitation of this technique is technical experience is required in drilling the thin shell of bone separating the optic foramen at the orbital apex from the cavernous segment of the internal carotid artery.

The Blurring and headache receded on day two of the surgery in our patient. Galbraith and Sullivan reported success in four cases of benign intracranial hypertension but the surgical success was described mainly in terms of papilloedema resolution.¹⁵

Kilpatrick et al described symptomatic relief post optic nerve fenestration in their 14 patients of benign intracranial hypertension, but the surgical indication was progressive visual failure in only six cases.¹⁶

Ventriculoperitoneal shunts for medically resistant benign intracranial hypertension have been reported to have shown improvements in headache, stabilising vision and/or reduction in papilloedema in paediatric population.¹⁷

CONCLUSION

Endoscopic trans-sphenoidal optic nerve sheath fenestration (ONSF) is an effective surgical technique to preserve vision in patients with progressive optic neuropathy due to papilledema caused by idiopathic intracranial hypertension (IIH).

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Conflict of interest: None declared

Ethical approval: Not required

REFERENCES

1. Degnan AJ, Levy LM. Pseudotumor cerebri: brief review of clinical syndrome and imaging findings. *AJNR Am J Neuroradiol.* 2011;32(11):1986-93.
2. Schirmer CM, Hedges TR. Mechanisms of visual loss in papilledema. *Neurosurg Focus.* 2007;23(5):E5.
3. Babikan P, Corbett J, Bell W. Idiopathic intracranial hypertension in children: the Iowa experience. *J Child Neurol.* 1994;9:144-9.
4. Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. *Neurol.* 2002;59:1492-5.
5. Bialer OY, Rueda MP, Bruce BB, Newman NJ, Biousse V, Saindane AM. Meningoceles in idiopathic intracranial hypertension. *AJR Am J Roentgenol.* 2014;202(3):608-13.
6. Leach JL, Fortuna RB, Jones BV, Gaskill-Shipley MF. Imaging of cerebral venous thrombosis: current techniques, spectrum of findings, and diagnostic pitfalls. *Radiographics.* 2006;26(Suppl 1):S19-41.
7. Fraser C, Plant GT. The syndrome of pseudotumor cerebri and idiopathic intracranial hypertension. *Curr Opin Neurol.* 2011;24:12-7.
8. DeWecker L. On incision of the optic nerve in cases of neuroretinitis. *Int Ophthalmol Cong Rep.* 1872;4:11-4.
9. Hamed LM, Tse DT, Glaser JS, Byrne SF, Schatz NJ. Neuroimaging of the optic nerve after fenestration for management of pseudotumor cerebri. *Arch Ophthalmol.* 1992;110(5):636-9.
10. Seiff SR, Shah A. Model for the mechanism of optic nerve sheath fenestration. *Arch Ophthalmol.* 1990;108(9):1326-9.
11. Mukherjee N, El-Dairi MA, Tariq Bhatti M. Optic nerve sheath fenestration-indications and techniques. *US Ophthalmic Rev.* 2013;6(2):125-31.
12. Feldon SE. Visual outcomes comparing surgical techniques for management of severe idiopathic intracranial hypertension. *Neurosurg Focus.* 2007;23(5):E6.
13. Lee AG, Patrinely JR, Edmond JC. Optic nerve sheath decompression in pediatric pseudotumor cerebri. *Ophthalmic Surg Lasers.* 1998;29(6):514-7.
14. Nithyanandam S, Manayath GJ, Battu RR. Optic nerve sheath decompression for visual loss in intracranial hypertension: Report from a tertiary care centre in South India. *Indian J Ophthalmol.* 2008;56:115-20.
15. Galbraith JEK, Sullivan JH. Decompression of the perioptic meninges for relief of papilloedema. *Am J Ophthalmol.* 1973;76:687-92.
16. Kilpatrick CJ, Kaufman DV, Galbraith JE, King JO. Optic nerve decompression in benign intracranial hypertension. *Clin Exp Neurol.* 1981;18:161-8.
17. Heyman J, Ved R, Amato-Watkins A, Bhatti I, Te Water NJ, Gibbon F, et al. Outcomes of ventriculoperitoneal shunt insertion in the management of idiopathic intracranial hypertension in children. *Childs Nerv Syst.* 2017;33(8):1309-15.

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