Pakistan Journal of Neurological Sciences (PJNS)

Volume 12 | Issue 1 Article 11

3-2017

Intracranial hypertension and optic nerve sheath fenestration

Irfan Jeeva
Aga Khan University Hospital, irfan.jeeva@aku.edu

INTRACRANIAL HYPERTENSION AND OPTIC NERVE SHEATH FENESTRATION

Irfan Jeeva[1]
[1]*Assistant Professor Aga Khan University Hospital*

Correspondence to: *Irfan Jeeva, Professor of Ophthalmology. Email: irfan.jeeva@aku.edu*

Date of submission: *June 18, 2016* ***Date of revision:*** *December 22, 2016* ***Date of acceptance:*** *December 29, 2016*

ABSTRACT

Intracranial Hypertension is a neurological condition secondary to raised intracranial pressure in the absence of identifiable intracranial pathology and with normal cerebrospinal fluid composition. It can be a challenging condition to manage. One of the treatment modalities, optic nerve sheath fenestration is an important, yet under utilized mode of treatment, particularly in patients with visual symptoms.

Key Words:
Intracranial hypertension, Cerebrospinal fluid, Papilledema , Optic nerve sheath fenestration

INTRODUCTION:

Intracranial Hypertension (IIH) is a neurological disorder characterized by raised cerebrospinal fluid pressure without identifiable intracranial pathology and with normal cerebrospinal fluid (CSF) composition.[1] This condition usually affects overweight women of childbearing age but can affect either gender at any age. The incidence is 0.9-2.2 per 100,000 in the general population rising to 19.3-21.3 per 100,000 in obese women of childbearing age.[1]

PATHOGENESIS:

The pathogenesis is unclear. Various factors including obesity, sex hormone, prothombotic factors and altered CSF dynamics have been suggested for the same.[2]
Obesity has long been associated with IIH. In a prospective study of 34 patients with IIH, Rowe[3] showed 94% of their patients to be overweight (BMI >26kg/m^2) and 73% obese (BMI >30 kg/m^2). Obese females between the ages 16-24 were 17 times more likely to develop IIH than their age-matched controls.[3]

PRESENTATION:

Patients with IIH can present with visual and non-visual symptoms. In a prospective study of 50 patients, Wall and George[4] reported headache in 94% of their patients, transient visual obscurations in 68%, intracranial noise in 58%, photopsia in 54% and retrobulbar pain in 44% of patients. Papilledema is considered to be the cardinal sign of IIH. In the absence of papilledema, obesity and pulsatile tinnitus can suggest high CSF pressure in a patient with chronic headaches. Wall and George[4] also reported impairment of visual acuity in 13% of their patients' initial visits and of visual field abnormality in 90% of their patients on perimetry. Typical visual field defects were blind spot enlargement, nasal defect, arcuate scotoma and generalized field constriction.

Friedman[5] described an updated criterion for diagnosing IIH (Table 1). The diagnosis of IIH requires presence of papilledema (Table 2 and 3), appropriate brain imaging to exclude intracranial abnormalities and a lumbar puncture to document raised intracranial pressure (>25cm H_2O) and normal CSF constituents.[5] Magnetic resonance imaging (MRI) is considered better than a computed tomography (CT) scan, however, in atypical cases magnetic resonance venography (MRV) along with MRI may be required. MRI may reveal signs of empty sella turcica, flattening of posterior globe and buckling of optic nerve. Ventricular size should be normal for the patient's age, however slit-like ventricle may be seen in IIH.[6]

IIH is a diagnosis of exclusion and conditions mimicking IIH should be excluded (Table 4). Various conditions are associated with IIH and need to be considered in the workup of the patient (Table 5)[7]. Bababeyge described two non-obese patients on minocycline who developed visual loss secondary to papilledema.[8]

Table 1. Modified Criteria for Diagnosis IIH [Friedman & Jacobson]

Criteria for diagnosing idiopathic intracranial hypertension
• Symptoms, if present, are only those of generalized intracranial hypertension or papilledema
• Signs, if present, are only those of generalized intracranial hypertension or papilledema
• Elevated CSF pressure demonstrated on lumbar puncture in the lateral decubitus position
• Normal CSF constituents
• No evidence of space occupying lesion or venous sinus thrombosis on imaging
• No other explanation for the raised CSF pressure

Table 2: Feature of papilledema

Features of papilledema
• Peripapillary nerve fiber layer edema obscuring the retinal blood vessels
• Absent spontaneous venous pulsations
• Distended retinal veins
• Peripapillary haemorrhages and exudates
• Peripapillary retinal or choroidal folds
• Nerve fiber layer infarcts

Table 3: Grading of papilledema

Stage	Description
0	Normal optic disc. Radial nerve fiber layers without nerve fiber layertortuosity. Blurring of nasal, superior, inferior margins inversely proportional to disc size. Greater the size, less the blurring.
1	C-shaped halo. Normal temporal margin. Concentric or radial retrochoroidal folds.
2	Circumferential halo. Nasal border elevation.
3	Obscuration of one or more segment of a major blood vessel.
4	Total obscuration on the disc of a segment of a major blood vessel. Elevation of the entire nerve head, including the cup.
5	Partial obscuration of all the blood vessels on the optic disc and total obscuration of at least one major blood vessel. Dome shape protrusion, representing anterior expansion of the optic nerve head.

Table 4: Differential Diagnosis

Important differential diagnosis of Idiopathic intracranial hypertension
Dural venous sinus thrombosis
Hypertensive retinopathy
Optic disc anomalies (disc drusen and tilted optic disc)
Intracranial hypertension due to medication or a disease

Table 5: List of systemic diseases and medications associated with intracranial hypertension

Systemic diseases and medications associated with intracranial hypertension	
Medication	**Systemic disease**
Tetracycline	Systemic Lupus Erythematosis
Minocycline	Behcet's disease
Doxycycline	Polycystic ovarian disease
Steroids	Hypothyroidism
Steroid withdrawal	
Vitamin A	

TREATMENT:

Treatment of IIH includes weight reduction, medical management, elimination of associated factors, serial lumbar puncture and surgical intervention

NON-SURGICAL TREATMENT

Johnson[9] observed that at least 6% of weight loss could result in reduction of ICP with accompanying resolution of papilledema and visual field dysfunction. Surgically induced weight loss has shown similar results.[7]

The mainstay of IIH medical management is acetazolamide, a carbonic anhydrase inhibitor that reduces CSF production by decreasing sodium ion transport across the choroidal epithelium. The starting dose is 250 mg twice daily to ensure tolerance, increasing to a maintenance dose of 1-2 g per day. Patients intolerant to acetazolamide can be treated with furosemide (loop diuretic) an ACE inhibitor or topiramate an anti-convulsion drug. IIH symptoms may resolve after lumbar puncture.[7]

SURGICAL TREATMENT

The surgical options are optic nerve sheath fenestration (ONSF) or CSF diversion procedures such as lumboperitoneal shunt (LPS) or ventriculoperitoneal shunts (VPS).

Optic nerve sheath fenestration (ONSF) was first described by DeWecker [10] to allow the flow of CSF from

the sub-arachnoid space to the orbital space. Almost a century later, in 1964, Hayreh [11] refined the technique of ONSF that resulted in the resolution of papilledema. This led to the theory that ONSF could be the preferred surgical option when headache is not the predominant symptom.[12] CSF shunting is considered to be the procedure of choice when headache is the predominant symptom.[13] However, studies have shown significant reduction of the symptoms of headache after ONSF, which may suggest the role of ONSF as an alternative, for patients with headaches as the predominant symptom.[14]

Banta and Ferris [15] described the effectiveness of ONSF in a case series involving 158 eyes by demonstrating improvement in the visual acuity of 94% of the eyes post surgery. In addition, 88% showed stabilization or improvement in the visual field. One eye suffered visually significant complication. The study reported less favorable outcome for the symptoms of headache. In this study, 61 patients presented with headache and only 31% reported improvement in their headache post ONSF.

A study by Chandrasekaran et al[16], on 51 eyes of 32 patients demonstrated that patients with mild visual field defect improved or stabilized post ONSF, while patients with severe visual field defect were found to stabilize post ONSF. Also younger patients showed more favorable result in this study. In the study it was found overall complication rate of 15.6%; all being self-limiting, three patients had diplopia, two had anisocoria and one had disc hemorrhage. All procedures were approached by medial subconjunctival approach and performed by the same surgeon. Plotnik and Kosmorsky[17] reported a complication rate of 40%, which included temporary motility disorder (29%), pupillary dysfunction (11%) and vascular complication (11%): comprising of two central retinal artery occlusion, one transient outer retinal ischemia and one superotemporal branch retinal artery occlusion.

Sergott[18] studied the benefit of ONSF in patients with prior LPS placement. The study enrolled 23 patients with previous multiple LPS; visual improvement was demonstrated in six patients after ONSF. This suggested that despite functional CSF shunt, ONSF could be beneficial in patients with progressive visual deterioration, or in the event of failure of the shunt.

In a retrospective review of 62 eyes by Alsuhaibani et al[19] found an additional benefit of unilateral ONSF to reduce the disc swelling in the contralateral un-operated eye. The reduced disc swelling in the contralateral eye continued improving with greatest effect seen 12 months post ONSF, corresponding with improvement in the visual function.

In general, ONSF is an effective long-term procedure for visual loss associated with IIH. Multiple studies have reported that majority of complication are often transient and resolve without long-term effects.

ONSF is considered to be the better option in patients with visual loss in IIH, while CSF shunting is the favorable option in patients with refractory headache. The two commonly performed CSF shunting procedures, ventricular peritoneal shunt (VPS) and Lumboperitoneal shunt (LPS), are considered to be almost similar in their outcome. Tarnaris et al[20], in a retrospective review of 34 patients with 63 shunts found no significant difference in visual outcome or headache between the two groups; VPS and LPS, however, headache improved more than visual disturbance for both the groups. The study also reported 20.5% of the patients had shunt procedure related complication with 35% requiring shunt revision. Abubaker 12 similarly found better outcomes with headache compared to visual disturbance post CSF shunting procedure and also noted LPS to have lower failure rate but higher revision rate. El-Saadany[21], in the retrospective review of 22 patients, found patients showed improvement in both headache (19/22) and papilledema (16/22). Moreover, the study reported 9% shunt infection rate, 27% shunt obstruction and 13% shunt over drainage. A 10-year retrospective review of 53 patients by Sinclair [22] found an overall improvement in the visual symptoms post-shunting but noted headache persisted in 79% of post-LPS patients. The review also noted a high rate of shunt complications, shunt revisions, and persistent post-shunt headache. Karabatsou[23] et al retrospectively reviewed 21 post LPS patients. The study highlighted[7] shunt related infections, 7 tonsillar herniations, 17 shunt migrations with 18 out of 22 patients requiring a total of 63 shunt revision procedures.

SURGICAL TECHNIQUE OF ONSF

Although many techniques have been described in literature, the preferred method for the author is lid crease approach. Other commonly used methods for access to the nerve are via medial orbitotomy and lateral orbitotomy.

Medial orbitotomy involves an approach to the optic nerve without cutting the skin. The medial rectus is detached and the optic nerve is identified. Once in view,

the optic nerve sheath is fenestrated to release the fluid. There is a risk of ocular muscle palsy with this surgery. Lateral orbitotomy approach involves skin incision, removal of the lateral orbital wall, giving a wide access to the optic nerve. However, it is a longer surgical procedure with a possible risk of damage to the ciliary body. The lid crease approach is gaining favor, as it does not involve bone removal or muscle detachment but the learning curve is considerably higher in this approach.[13]

SOCIETAL IMPACT OF IDIOPATHIC INTRACRANIAL HYPERTENSION:

The societal impact of this condition is high and often goes unrecognized. According to the intracranial hypertension registry in the USA, in 2007 the total cost of IIH was estimated to be 444 million USD, of which 232 million USD is attributable to direct medical and surgical expense.[24]

CONCLUSION

IIH can cause visual loss. Medical management is the first line of treatment in IIH. In refractory cases and in patients presenting with visual loss, optic nerve sheath fenestration is an important and viable treatment option to prevent sight loss in this challenging neuro-ophthalmological condition.

REFERENCE:

1. Dhungana S, Sharrack B, Woodroofe N. Idiopathic Intracranial Hypertension. Acta Neurol Scand. 2010:121(2):71-82
2. Backhouse O, Johnson M, Jamieson DR et al. Familial thrombophilia and idiopathic intracranial hypertension. Neuroophthalmology 2001;25:135–41.
3. Rowe FJ, Sarkies NJ. The relationship between obesity and idiopathic intracranial hypertension. Int J Obes Relat Metab Disord 1999;23:54–9.
4. Wall M, George D. Idiopathic intracranial hypertension (pseudotumour cerebri), a prospective study of 50 patients. Brain 1991;114:155–80
5. Friedman D I, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. Neurology. 2002;59(10):1492-5.
6. Brodsky MC, Vaphiades M. Magnetic resonance imaging in pseudotumor cerebri. Ophthalmology 1998;105:1686–93.
7. Mathews MK, Sergott RC, Savino PJ. Pseudotumor cerebri. Curr Opin Ophthalmol. 2003 Dec;14(6):364-70.
8. Bababeygy SR, Repka MX, Subramanian PS. Minocycline associated pseudotumor cerebri with severe papilledema J Ophthalmol. 2009;2009:1-5.
9. Johnson LN, Krohel GB, Madsen RW, March GA Jr. The role of weight loss and acetazolamide in the treatment of idiopathic intracranial hypertension (pseudotumor cerebri). Ophthalmology 1998;105: 2313–7.
10. DeWecker L. On incision of the optic nerve in cases of neuroretinitis. Rep Int Ophthalmol Congr. 1872;4:11–4.
11. Hayreh SS.Pathogenesis of oedema of the optic disc (Papilloedema). A preliminary report. Br J Ophthalmol 1964;48:522-43.
12. Abubaker K, Ali Z, Raza K, Bolger C, Rawluk D, O'Brien D. Idiopathic intracranial hypertension: lumboperitoneal shunt versus ventriculoperitoneal shunts – case serried and literature review. British Journal of Neurosurgery, February 2011;25:94-9.
13. Prabhakaran VC, Selva D. Vertical lid split approach for optic nerve sheath decompression. Indian J Ophthalmol 2009;57:305-6.
14. Brazis P. Clinical review: The surgical treatment of idiopathic pseudotumor cerebri (idiopathic intracranial hypertension). Cephalgia 2008;28:1361-73
15. Banta JT, Farris BK. Pseudotumor cerebri and optic nerve sheath decompression. Ophthalmology 2000;107:1907-12.
16. Chandrasekaran S, McCluskey P, Minassian D, Assaad N. Visual outcomes for optic nerve sheath fenestration in pseudotumor cerebri and related conditions. Clin Experiment Ophthalmol 2006;34:661-5.
17. Plotnik JL, Kosmorsky GS. Operative complications of optic nerve sheath decompression. Ophthalmology 1993;100:683-90.
18. Sergott RC, Savino PJ, Bosley TM. Modified optic nerve sheath decompression provides long-term visual improvement for pseudotumor cerebri. Arch Ophthalmol 1988;106:1384-90.
19. Alsuhaibani AH, Carter KD, Nerad JA, Lee AG. Effect of optic nerve sheath fenestration on papilledema of the operated and the contralateral nonoperated eyes in idiopathic intracranial hypertension. Ophthalmology 2011;118:412-4.
20. TarnarisA,TomaAK,WatkinsLD,KitchenND.Istheread ifference in outcomes of patients with idiopathic intracranial hypertension with the cho\ cerebrospinal fluid diversion site: A single centre

experience. Clin Neurol Neurosurg 2011;113:477-9.
21. El-Saadany WF, Farhoud A, Zidan I. Lumboperitoneal shunt for idiopathic intracranial hypertension: Patients' selection and outcome. Neurosurg Rev 2012;35:239-43.
22. Sinclair AJ, Kuruvath S, Sen D, Nightingale PG, Burdon MA, Flint G. Is cerebrospinal fluid shunting in idiopathic intracranial hypertension worthwhile? A 10-year review. Cephalalgia 2011;31:1627-33.
23. Karabatsou K, Quigley G, Buxton N, Foy P, Mallucci C. Lumboperitoneal shunts: Are the complications acceptable? Acta Neurochir (Wien) 2004;146:1193-7.
24. Friesner D "The Costs of a Quiet Disorder: Direct and Indirect Costs of Idiopathic Intracranial Hypertension," Obesity Reviews, 2011;12:e372-e380

Conflict of interest: Author declares no conflict of interest.
Funding disclosure: Nil

Author's contribution:
Irfan Jeeva; Study concept and design, protocol writing, data collection, data analysis, manuscript writing, manuscript review