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Ophthalomology

REVIEW ON OPHTHALMIC MANIFESTATIONS OF IDIOPATHIC INTRACRANIAL HYPERTENSION

Dr Dheerendra Singh MBBS, MS, FICO(U.K), FASG (Phacoemulsification and Medical Retina

Dr Manisha Singh* MBBS, MS, FVRS, Vitreo-retina Consultant, Ajwani Eyecare centre, Bhopal.*Corresponding Author

ABSTRACT Idiopathic intracranial hypertension (IIH) also known as Benign Intracranial Hypertension or Pseudotumor Cerebri is an idiopathic condition that is associated with increased intracranial pressure of unknown origin. It is usually associated with headaches, papilloedema, vision changes, or pulsatile tinnitus in the setting of normal imaging and cerebrospinal fluid (CSF) studies. It is concurrent with obesity with in females between 20-44 years of age. visual disturbances and patient may even progress to blindness if untreated. Serial perimetry and optic disc grading or fundus photography plays a crucial role in appropriate management. Medical management and weight loss are beneficial in improving both quality of life and ICP in obese patients with IIH

KEYWORDS : Benign Intracranial Hypertension, Idiopathic intracranial hypertension (IIH), Pseudotumor Cerebri, papilloedema

INTRODUCTION

Idiopathic intracranial hypertension (IIH) also known as Benign Intracranial Hypertension or Pseudotumor Cerebri is an idiopathic condition that is associated with increased intracranial pressure of unknown origin. [1] The first case of IIH was reported by Quincke in 1897, and the term Benign Intracranial Hypertension was coined by Foley in 1955. [2] It is characterized by severe headache, pulsatile tinnitus, and transient visual obscurations with raised intracranial pressure (ICP) on lumbar puncture (LP) in the absence of an intracranial mass or venous sinus thrombosis. Papilloedema is a key neuro-ophthalmologic feature and its presence or absence is a major metric of both disease severity and treatment success. Visual loss is often the primary concern in these patients; the disease frequently results in severe impairments in quality of life as well. [3] Medical therapies may be effective in mitigating the risk of visual loss but frequently result in undesirable side effects that add to the symptom burden. Medical management and weight loss have been reported to be beneficial in improving both quality of life and ICP in obese patients with IIH.[4]

Epidemiology

Annual incidence is 0.9/100000 persons with 3.5/100000 in females between 15 to 44 years of age. It is concurrent with obesity with the incidence increasing upto 19 per 100000 in females between 20-44 years of age that are 20% or more over the ideal weight.[5] Obesity is known to be the most important contributing factor as more than 90% of patients are obese and over 90% are women of childbearing age with asymptomatic elevated intracranial pressure that may persist for year. [6] The peak incidence is in 3rd decade of life.[7]

Pathophysiology

Earliest proposed mechanism was cerebral edema, however, the raised ICP was not associated with alteration of alertness, cognitive deficits, or focal neurologic symptoms indicative of cerebral edema.

Stenosis of the transverse venous sinuses distal section is a different suggested process that may reduce CSF absorption and cause cerebral venous hypertension. The venous sinus stenosis may be an accidental finding, the secondary cause of the raised ICP, or the major cause of the elevated ICP, according to the most recent literature.

Raised intraabdominal pressure due to obesity also increases heart filling pressure that may lead to increase in intracranial venous pressure and IIH. This theory, fails to explain its occurrence in the non-obese population.

Vitamin A may also play some role pertaining to higher amounts of vitamin A, retinol, and retinol binding protein found in the serum and CSF of IIH patients.

Moreover, it has been hypothesized that a micro thrombosis in the sagittal sinus that is too small to be observed on neuroimaging may be preventing CSF from entering the arachnoid granulations. The absence of hydrocephalus, which is typically associated with poor CSF absorption or overproduction, contradicts this idea.

Clinical Features

- Headache (94%) worse in the morning aggravated by Valsalva's maneuver.[8]
- Transient visual obscurations (68%) usually preceded by headache caused by transient ischemia of the optic nerve head related to increased tissue pressure.
- Tinnitus synchronous with pulse rate (58%).
- Photopsia (54%)
- Retrobulbar pain (44%)
- Diplopia (38%)5{cranial nerves II, VI, VII most commonly affected}[9]
- Visual loss (30%).[10]

Pseudotumour cerebri may be associated with:-

(a) Obstruction /impairment of cerebral venous drainage (tumors, septic thrombi, radical neck dissection).

(b) Endocrine and metabolic dysfunction

- Elevated estrogen levels
- Aldosterone excess (affect the mineralocorticoid receptor of the choroid plexus that leads to increased CSF production)
- Pregnancy (esp.2nd and 5th month due to decreased corticoids and increased estrogen).[6]
- Hypoparathyroidism and hypocalcemia (interferes with transport of CSF through arachnoid granulation).

(c) Exogenous agents

- Systemic corticosteroids (suppression of adrenal cortex)
- Antibiotics(Tetracycline, Nalidixic acid)
- Anti-inflammatory drugs(Indomethacin, Ketoprofen)
- Vitamin A overdose (1 lakh units per day for a few months)
- Lead encephalopathy (causing cerebral edema and increased intracranial tension).

(d) Systemic illnesses

- Meningitis, Encephalitis (blockage of ventricular system)
- Status Epilepticus (cerebral hypoxia and cerebral edema)
- Vascular hypertension
- Thrombocytopenic purpura
- Chronic respiratory insufficiency.

(e) Familial Benign Intracranial Hypertension.[11]

INVESTIGATIONS

1) Fundus examination

Papilledema or optic disc edema [Figure 1] due to increased intracranial pressure is the most important sign of benign intracranial hypertension that may even lead to visual loss. The higher the grade of the papilledema, the worse the visual loss.[12]

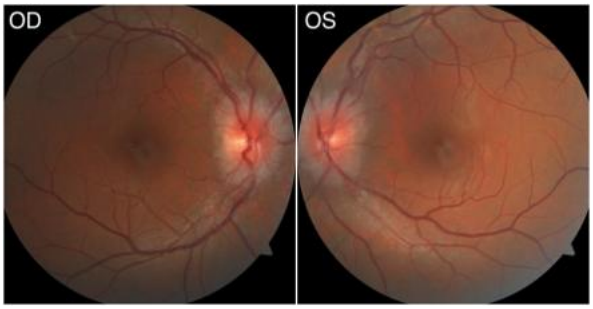

Figure 1- fundus Findings S/o B/l Disc Edema (papilledema)

2) Perimetry

Inferior nasal step defect is the earliest visual field defect followed by peripheral nasal loss. Arcuate defects may appear next followed by a gradual depression of the entire field, most pronounced peripherally along with blind spot enlargement [Figure 2] which is usually present.[13]

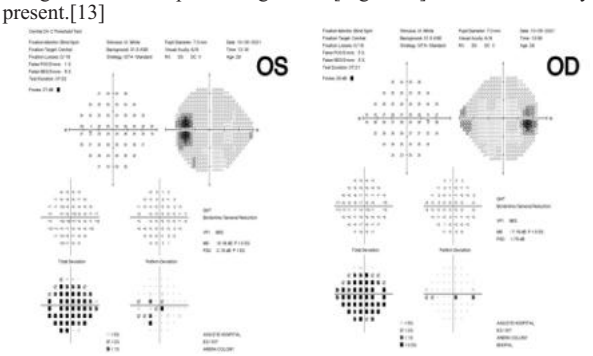

Figure 2 : Visual Fields In Both Eyes S/o Enlarged Blind Spot

3) Optical Coherence Tomography

Important for both diagnosis as well as monitoring the condition. There is an upward deflection of Bruch's membrane. Also, elevated optic nerve head with smooth internal contour and subretinal hyporeflective space (SHYPS) with recumbent "lazy V" pattern can be seen with 3-D display of optic nerve [Figure 3] being very helpful.[14]

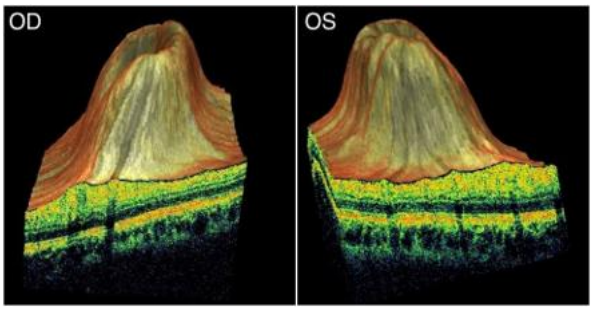

Figure 3: Elevation Of Nerve Fiber On 3d Display S/o B/l Disc Edema

4) Neuroimaging

- MRI Brain and Orbit may reveal-
- Distension of perioptic subarachnoid space
- Partial empty sella
- Flattening of posterior sclera
- Vertical tortuosity and elongation of orbital Optic Nerve
- Bilateral transverse sinus stenosis.[1,15]

5) Diagnostic Lumbar Puncture

For CSF opening pressure.[1-6]

Pathological entities like venous obstruction (e.g. venous sinus thrombosis) and leptomeningeal diseases (e.g. meningitis) can mimic these intracranial findings even without the mass effect. Therefore, CT or MR venography and lumbar puncture are part of the routine workup of suspected IIH.

DIAGNOSIS

Diagnostic criteria were first given by Walter Dandy in 1937, later modified by Smith in 1985 and by Digre and Corbett in 2001.

Modified Dandy criteria

- Signs and symptoms of raised intracranial pressure(headache, nausea, vomiting, papilledema)
- No localizing findings on neurological examination(except false localizing signs such as CN VI or VII palsy)
- Awake and alert patient
- Increased CSF opening pressure of >25 cm or 250 mm of water measured in left lateral decubitus position
- Normal CSF cytological and chemical composition
- Normal CT/MRI/MR Venography findings and normal neurological examination except papilledema with no evidence of dural sinus thrombosis
- No other cause of increased intracranial, pressure found.[1-4]

Revised Friedman criteria

New diagnostic criteria were proposed in 2013 and are also commonly used [16]. The original terminology was for pseudotumor cerebri syndrome but the term idiopathic intracranial hypertension has since supplanted it.

The criteria place patients into one of four diagnostic subgroups:

- Definite idiopathic intracranial hypertension: opening pressure ≥25 cm CSF (H_2O) and papilledema
- Probable idiopathic intracranial hypertension: opening pressure <25 cm CSF and papilledema
- Definite idiopathic intracranial hypertension without papilledema: opening pressure ≥25 cm CSF and abducens nerve palsy
- Suggested idiopathic intracranial hypertension without papilledema: opening pressure ≥25 cm CSF and ≥3 out of 4 neuroimaging signs (empty sella, flattening of the posterior aspect of the globe, distension of the perioptic subarachnoid space, or transverse venous sinus stenosis)

These subgroups have several additional common requirements:

- Neurologic examination: normal except for cranial nerve abnormalities
- Neuroimaging: no hydrocephalus, mass, structural lesion, or abnormal meningeal enhancement on MRI (without and with contrast) and, if not female or obese, MRV (if MRI is unavailable or contraindicated, contrast-enhanced CT may be used)
- Laboratory: normal CSF composition

For children, the threshold opening pressure is 28 cm CSF rather than 25 cm CSF, unless the child is not sedated and not obese.

MANAGEMENT

(A)Conservative management

1) Weight loss

Weight loss in the range of 5-10% of total body weight can cause reversal of signs and symptoms. Too much body fat in chest and belly puts pressure on those areas, making it more difficult for blood from brain to get there. Fluid buildup in the brain can therefore raise risk of IIH [17]

2) Acetazolamide

Considered as treatment of choice. In adult patients, it is usually started at 1 g daily (250 mg QID or 500 mg BID), with a maximum recommended daily dose of 4 g. Adverse effects of acetazolamide include paraesthesias, lethargy and altered taste. Hypokalemia is an important adverse effect and hence, electrolytes must be monitored. Idiopathic Intracranial Hypertension Treatment Trial reports improved quality of life outcome with use of acetazolamide at 6 months.[18]

3) Furosemide

Lowers the intracranial pressure by causing diuresis and reducing sodium transport into the brain. Dose of 20 mg p.o. b.i.d. to a maximum of 40 mg p.o. t.i.d can be given. Potassium supplementation is given as needed.[19]

4) Topiramate

Acts by inhibiting carbonic anhydrase and can also suppress appetite. The dose of is from 25 mg to 50 mg bd with side effects like depression, cognitive slowing and potential teratogenic risks.[20]

5) NSAIDs

Paracetamol may be used for headache. Indomethacin has an advantage of reducing ICP.[21]

6) Corticosteroids
Short term course can also be used pre operatively before CSF shunting procedure.[22]

7) Therapeutic Lumbar Puncture
Has only a short-lived effect on CSF pressure with a return of pressure to pre-tap level after only 82 minutes.[23]

(B)Surgical Management
1) Subtemporal /Suboccipital decompression
Long term success rates are reported Complications include seizures, otorrhea, and subdural hematoma.

2) Optic nerve sheath decompression/fenestration
Preferred for patients with progressive visual loss with mild or easily controlled headaches.Has good efficacy.

3) CSF Shunting
Lumbar subarachnoid-peritoneal shunts, ventriculo-atrial, ventriculo-jugular and ventriculoperitoneal shunts can be done for a failed medical therapy or intractable headache and has been increasingly used nowadays. The extra cerebrospinal fluid is eliminated by the shunt [24]

4) Gastric exclusion surgery
Useful in treating co morbid conditions with obesity such as arterial hypertension, diabetes mellitus, and sleep apnea. Complications include major wound infection and stenosis at the gastro-jejunal anastomosis. Has been successful for morbid obese patients.[25]

5) Venous Sinus stenting
Novel method, tried in cases of transvers sinus stenosis and requires further study.[26]

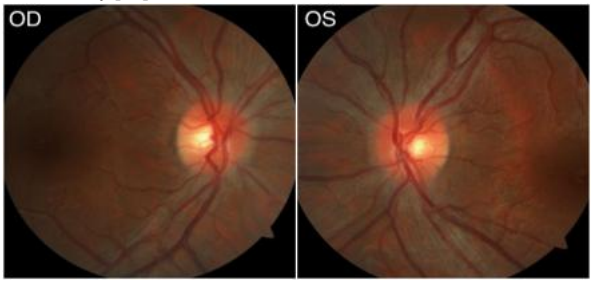

FIGURE 4 Post treatment fundus findings s/o decrease in disc edema

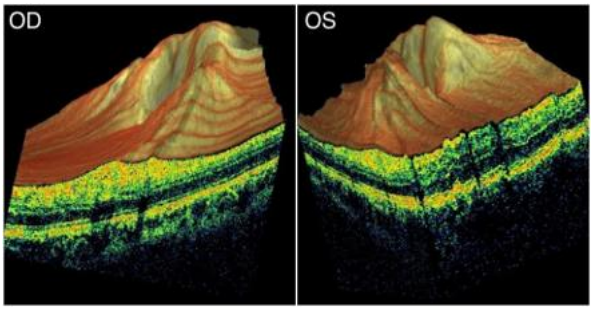

FIGURE 5- 3 D OCT - Post treatment Nerve fiber

CONCLUSIONS

Idiopathic intracranial hypertension is characterized by elevated CSF (intracranial) pressure of unknown origin. It is predominantly seen in women of childbearing age with complaints of severe headache, visual disturbances and patient may even progress to blindness if untreated. Serial perimetry and optic disc grading or fundus photography plays a crucial role in appropriate management. CT or MR venography and lumbar puncture are part of the routine workup of suspected IIH to rule out venous obstruction and leptomeningeal diseases. There is no evidence-based data to guide therapy, hence, there are still ongoing randomized double blind controlled treatment trials investigating diet and medical therapy.

REFERENCES:

1. Ambika S,Arjundas D,Noronha V,Anshuman.Clinical profile,evaluation,management and visual outcome of idiopathic intracranial hypertension in a neuro-opthalmology clinic of a tertiary referral ophthalmic center in India.Ann Indian Acad Neurol 2010;13(1):37-41.
2. Corbett JJ, Savino PJ, Thompson HS, et al. Visual loss in pseudotumor cerebri. Follow-up of 57 patients from five to 41 years and a profile of 14 patients with permanent severe visual loss. Arch Neurol. 1982;39:461–474.
3. Garner RM, Aldridge JB, Wolfe SQ, et al. Quality of life, need for retreatment, and the re-equilibration phenomenon after venous sinus stenting for idiopathic intracranial hypertension. J Neurointerventional Surg 2021; 13: 79–85.
4. Mollan SP, Mitchell JL, Ottridge RS, et al. Effectiveness of bariatric surgery vs community weight management intervention for the treatment of idiopathic intracranial hypertension: a randomized clinical trial. JAMA Neurol 2021; 78: 678–686.
5. Durcan FJ, Corbett JJ, Wall M. The incidence of pseudotumor cerebri. Population studies in Iowa and Louisiana. Arch Neurol. 1988;45:875–877.
6. Corbett JJ. The 1982 Silversides lecture. Problems in the diagnosis and treatment of pseudotumor cerebri. Can J Neurol Sci. 1983;10:221–229.
7. Wall M, George D. Idiopathic intracranial hypertension. A prospective study of 50 patients. Brain. 1991;114:155–180.
8. Wall M. The headache profile of idiopathic intracranial hypertension. Cephalalgia. 1990;10:331–335.
9. Cushing H. Strangulation of the nervi abducentes by lateral branches of the basilar artery in cases of brain tumor. Brain. 1910;33:204–235
10. Knight RS, Fielder AR, Firth JL. Benign intracranial hypertension: visual loss and optic nerve sheath fenestration. Journal of Neurology, Neurosurgery & Psychiatry. 1986 Mar 1;49(3):243-50.
11. Chen J, Wall M. Epidemiology and risk factors for idiopathic intracranial hypertension. International ophthalmology clinics. 2014;54(1).
12. Wall M, White WN. Asymmetric papilledema in idiopathic intracranial hypertension: prospective interocular comparison of sensory visual function. Invest Ophthalmol Vis Sci. 1998;39:134–142
13. Corbett JJ, Jacobson DM, Mauer RC, Thompson HS. Enlargement of the blind spot caused by papilledema. Am J Ophthalmol. 1988;105:261–265.
14. Lenworth N. Johnson,, Meredith L. Diehl, Chuck W. Hamm, Drew N. Sommerville,Gregory F. Petroski, et al. Differentiating Optic Disc Edema From Optic Nerve Head Drusen on Optical Coherence Tomography. Arch Ophthalmol 2009; 127(1):45-49.
15. Riggeal BD, Bruce BB, Saindane AM,Ridha MA, Kelly LP, Newman NJ, et al. Clinical course of idiopathic intracranial hypertension with transverse sinus stenosis. Neurology 2013; 80(3):289-95.
16. Friedman D, Liu G, Digre K. Revised Diagnostic Criteria for the Pseudotumor Cerebri Syndrome in Adults and Children. Neurology. 2013;81(13):1159-65.
17. 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;105:2313–2317.
18. Michael Wall. Update on idiopathic intracranial hypertension. Neurol Clin. 2017; 35(1):45-57
19. Pollay M, Fullenwider C, Roberts PA, Stevens FA. Effect of mannitol and furosemide on blood-brain osmotic gradient and intracranial pressure. J Neurosurg. 1983;59:945–950.
20. Celebisoy N, Gökçay F, Sirin H, Akyürekli O. Treatment of idiopathic intracranial hypertension: Topiramate vs acetazolamide, an open-label study. Acta Neurol Scand 2007; 116:322-7.
21. Sader N, Zeiler FA, Gillman LM, et al. indomethacin for control of iCP. Neurocrit Care 2015; 22:437–49.
22. Errquiq L,Benomar A,Aitbenhaddou E et al .Clinical and therapeutic aspects of benign intracranial hypertension .Rev Neurol 2004;160(12):1187-90.
23. Johnston I, Paterson A. Benign intracranial hypertension. II. CSF pressure and circulation. Brain. 1974;97:301–312.
24. Mukherjee N, Bhatti MT. Update on the surgical management of idiopathic intracranial hypertension. Current neurology and neuroscience reports. 2014 Mar 1;14(3):438.
25. Sugerman HJ, Felton WL, III, Sismanis A, Kellum JM, DeMaria EJ, Sugerman EL. Gastric surgery for pseudotumor cerebri associated with severe obesity. Ann Surg. 1999;229:634–640.
26. Friedman DI. Cerebral venous pressure, intra-abdominal pressure, and dural venous sinus stenting in idiopathic intracranial hypertension. J Neuroophthalmol. 2006;26:61–64.