Idiopathic Intracranial Hypertension (Iih) Revisited

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Abstract

Idiopathic intracranial hypertension (IIH) or Benign intracranial hypertension (BIH) is an uncommon disorder characterized by signs and symptoms of raised intracranial pressure of unknown cause and usually without any evidence of intracranial pathology. This condition typically affects obese women. The incidence of IIH is increasing with the rising prevalence of obesity. The most common symptoms are headache, nausea, vomiting, pulsatile tinnitus, transient episodes of visual loss, diplopia and other visual symptoms. Papilledema is a major clinical sign. Visual impairment is a serious complication that may not be recognized by the patients. As its presenting symptoms mimic those of a brain tumour, it's also called pseudotumor cerebri (PTC). This paper in brief reviews epidemiology, clinical manifestations, etiopathogenetic hypothesis, diagnostic challenges, and current treatments of IIH including medical, surgical, and interventional management of this condition. The efficacy and complications of surgical interventions like cerebrospinal fluid diversion, optic nerve sheath fenestration, and endovascular venous stenting reported in recent two to three decades have been summarized in brief.

Keywords: Intracranial; Hypertension; (Iih) Revisited.

Introduction

Idiopathic intracranial hypertension (IIH) or Benign intracranial hypertension (BIH) is acondition characterized by signs and symptoms of raised intracranial pressure of unknown cause. The most common symptoms are headache, nausea, vomiting, pulsatile tinnitus, transient episodes of visual loss, diplopia and other visual symptoms. Papilledema isa major clinical sign.[1] As its presenting symptoms mimic those of a brain tumour, it's also called pseudotumor cerebri (PTC).

The first case of IIH named "serous meningitis" was reported by the German physician Heinrich Quincke in 1893. Compatriot Max Nonnein in 1904 introduced term "pseudotumor cerebri", brain tumor mimicking condition. Walter Dandy in 1937 developed Diagnostic criteria for IIH and introduced sub temporal decompressive surgery. In 1955 renamed as "Benign intracranial hypertension to distinguish it from intracranial hypertension due to secondary causes. Name was revised in 1989 to "Idiopathic intracranial hypertension".

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Epidemiology and Risk Factors

The disorder may occur in all age groups, but is most common in young women between the ages of 20 and 50, especially those with obesity.[1] Globally obesity is a major risk factor with an increased risk of severe vision loss due to IIH. The approximate annual incidence in United States has been reported to be 0.9 to 1.0 per 100,000 in the general population, 1.6-3.5 per 100,000 in women and 7.9-20 per 100,000 in over weight women. The estimated annual incidence of IIH in Middle East countries is 2.02–2.2/100,000 in the general

population, which is higher than the Western statistics and the incidence of obesity as well as IIH is much lower in Asian countries.[2] Indian patients have an overall higher rate of obesity as per WHO criteria but shows low prevalence of obesity in IIH than the global prevalence.[3] There is no difference in incidence between males and females in children. There is no known proven genetic cause for IIH.[4]

It is more a diagnosis of exclusion when no possible causes are found. The condition is termed as " Secondary intracranial hypertension" if there is an underlying cause. Although the underlying mechanisms are not yet fully understood, certain systemic illnesses including Addison disease, anemia, systemic lupus erythematosus, obstructive sleep apnoea, hypothyroidism, Bechet's syndrome, polycystic ovary syndrome, coagulation disorders and certain medications like vitamin A, lithium, anabolic steroids, oral contraceptive pills, nalidixic acid cyclosporine have been associated.

Clinical Presentation

Usually clinical presentation of IIH is non-specific, commonly presenting symptoms are:

Headache: Seen in 92–94% of the cases, frontal, throbbing in nature, may be associated with vomiting, may also occur with retro-ocular pain. Worse in morning and by activities like coughing and sneezing that further increases ICT.[5]

Pulsatile Tinnitus: Diagnostic specific, unilateral or bilateral "whooshing" sound, synchronous with the pulse and exacerbated with positional changes.[1]

Visual Symptoms

Transient episodic vision loss, lasting for few seconds usually following changes in posture or with Valsalva manoeuvres.

Visual field disturbances, typically peripheral visual field with an inferonasal defect, arcuate defect, or severe visual field constriction.

Horizontal diplopia, due to unilateral or bilateral non-localizing sixth cranial nerve palsy. The increased pressure leads to compression and traction of the cranial nerves, commonly sixth nerve is involved and rarely third and fourth nerves are affected.[6]

Papilledema is the hallmark sign of IIH. Typically, bilateral and symmetric, unilateral or asymmetrical cases may also occur. The severity grading can be done by Frisen scale. Long standing papilledema cases leads to Optic atrophy.[7]

Nonspecific rare symptoms associated with IIH are numbness of the extremities, generalized weakness, loss of smell, and dysco-ordination.[1]

Choroidal compression across macula, choroidal neovascularisation, and retinal elevation around optic disc may be found on fundoscopic examination.

Pathophysiology

Despite of multiple hypotheses the cause and pathophysiology of IIH remain unclear. Different theories of pathological mechanisms of IIH are

Cerebraloedema: Cerebral oedema leads to IIH, is the earliest proposed pathological mechanism was quickly criticized because, the elevated ICP was not associated with other signs and symptoms typically seen with cerebral oedema.[7]

Venous Sinus Stenosis: Stenosis of the distal portion of the transverse venous sinusesresult in cerebral venous hypertension and impaired CSF absorption. Multiple studies demonstrated evidence of bilateral venous sinus stenosis in IIH patients. However, it is still not clear whether the venous sinus stenosis is the primary cause of the elevated ICP, secondary to the elevated ICP, or an incidental finding.[8]

Intraabdominal Pressure: elevated intracranial venous pressure and IIH in obese patients due to increased intraabdominal pressure alters cardiac filling pressure which further impedes venous return from the brain. This hypothesis failed to explain the incidence among the non-obese.[9]

Vitamin A: elevated serum and CSF vitamin A, retinol, and retinol binding protein levels reported in IIH patients. Implications of these findings remain unclear.[10]

Other proposed theories include micro thrombosis in the sagittal sinus which is blocking CSF absorption in the arachnoid granulations.[11]

Diagnosis

The diagnosis of IIH may be suspected on the basis of the history, complete ocular examination including a dilated fundus examination, visual field examination, and optic nerve photographs

and exclusion of secondary causes by diagnostic procedures including neuroimaging and Lumbar puncture.

Modified Dandy Criteria: With modifications of diagnostic criteria established by Dandy in 1937 for the diagnosis of IIH:[12]

1. Signs and symptoms of increased ICP include headaches, nausea, vomiting, transient visual obscurations, papilledema.
2. No localizing neurologic signs, except for unilateral or bilateral sixth cranial nerve palsy.
3. CSF opening pressure >25 cm H2O with normal CSF composition.
4. No evidence of hydrocephalus, mass, structural, or vascular lesion (including venous sinus thrombosis) on imaging.
5. Not identified other cause for increased ICP.

Differential Diagnosis

Disc oedema secondary to elevated intracranial pressure termed as papilledema, Other causes of papilledema must be considered for the diagnosis are:[13]

- Intracranial mass lesions,
- Obstructive hydrocephalus,
- Conditions like venous sinus thrombosis, jugular vein compression, neck surgery obstructing the venous outflow.
- Increased CSF production conditions like choroid plexus papilloma.
- Decreased CSF absorption conditions e.g., arachnoid granulation adhesions after bacterial or other infectious meningitis, s haemorrhage

Differential diagnosis should also include: Pseudo papilledema, Papillitis, optic neuropathies, Orbital optic nerve tumours.

Diagnostic Procedures

Computed tomography (CT/CAT) or magnetic resonance imaging (MRI) detect any mass lesions. These scans may be normal or flattening of the pituitary gland due to increased pressure, the «emptysella sign" may be seen. MR venography (MRV) is usually indicated in atypical cases to exclude the possibility of venous obstruction or cerebral venous sinus thrombosis.[14]

Other Neuroimaging findings suggestive of IIH but also seen with other causes of increased ICPare flattening of the posterior pole, distension of peri optic subarachnoid space, vertical tortuosity of the orbital optic nerve, intraocular protrusion of the prelaminar optic nerve, stenosis of transverse cerebral venous sinuses.

Lumbar punctureto measure the opening pressure and to obtain cerebrospinal fluid (CSF) to exclude alternative diagnoses. Elevated opening pressure greater than 25 cm H2O with the patient lying in the lateral decubitus position considered for diagnostic criteria. The CSF examined to rule-out inflammation, tumour cells, and infection. In IIH, CSF shows normal or low protein level, normal glucose levels and a normal cell count.[15]

Management

General Treatment

The primary goal of treatment is to alleviate symptoms of ICP and preserve vision. Diagnostic lumbar puncture may relive symptoms, in some cases this is sufficient to control the symptoms, can be repeated to control the ICP urgently in rapidly vision deteriorating patients. Repeated lumbar punctures present a danger of introducing spinal infections.

Obese patients are encouraged to lose weight. 5-10% weight loss among obese/overweight patients has been found to be related to improved symptoms and signs. Preservation of weight loss and avoidance of weight fluctuation minimises the risk of recurrence. Bariatric surgery may be an option for morbidly obese. Simultaneous other modalities of treatment should be used on the long-term.[16]

Medical Therapy

For patients with mild to moderate disease usually medical therapy is considered.

Carbonic Anhydrase Inhibitor: Acetazolamide, the first-line medical treatment, by inhibiting the enzyme carbonic anhydrase, reduces the rate of CSF production. It can cause muscle weakness and tingling sensation in the fingers, symptoms of hypokalaemia and is contraindicated in pregnancy.

If Acetazolamide is inefficient or not tolerated, it may be combined with or substituted by anticonvulsant Topiramate, a weak carbonic anhydrase inhibitor, it has similar efficacy to acetazolamide for visual symptoms. Diuretic, Furosemide may also be used, but is not as effective at reducing ICP.[17]

Convention alanalgesic agent like paracetamol, antidepressant amitriptylinein low dose have some additional benefit for pain.

The use of steroids is controversial to reduce the ICP, these may be used in severe papilledema, but otherwise their use is discouraged.

Surgical Management

In patients with refractory headaches, rapidly progressive visual field loss and when medical therapy failed to prevent progressive visual loss, surgical management may be considered.

Indicationsfor Surgery: Severe visual loss, worsening of visual field defect, development of a new visual-field defect, Psychosocial reasons include non-compliance to medication, inability to perform visual field studies, refractory headache

Surgical Procedures

Optic Nerve Sheath Fenestration: Preferred surgical procedurein patients with papilledema, severe vision loss but no or minimal ICP symptoms. The procedure creates an outlet for continuous CSF drainage by giving incisions in the anterior dura covering of the optic nerve sheath. Axoplasmic flow in the optic nerve is restored and the CSF no longer distends the sheath. Safest procedure for renal failure patients and for pregnant.[18]

Csf Shunting: Preferred and most beneficial surgical procedurein patients with vision loss and symptoms of raised ICP for rapid reduction of ICP. Ventriculo -peritoneal (VP) and lumbo-peritoneal (LP) are the two types. Due to lower complication rate, VP shunting is the preferred method. Shunt obstruction, shunt migration, intracranial hypotension, and tonsillar herniation are the complications of CSF shunting. shunt revisions and multiple revisions may be needed in many patients.[18]

Venous Sinus Stenting (Vss): Preferred procedure in patients with venous sinus stenosis. It involves identification of the stenotic area on MRV, establishment of pressure gradient typically ≥ 10mmHg and stent placement. VSS decreases cerebral venous pressure, leading to increased CSF absorption and consequent decrease ICP. Stent migration, venous sinus perforation, in-stent thrombosis, subdural haemorrhage are the associated serious complications.[18-20]

Surgical intervention should be the earlier option in the paediatric patients.

Prognosis

There are no sufficient studies that have evaluated the natural history of the disease, the course may vary from weeks to years. Usually course of the disease is improvement and/or disease stabilization following treatment. Persistent visual field defects, disc oedema or elevated opening pressures on lumbar puncture may be demonstrated in many patients. Male gender, black race, morbid obesity, anemia, obstructive sleep apnoea are the worse visual outcome factors. 8 to 38% of patients shows recurrences, Weight gain is the main factor associated with disease recurrence.

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