IDIOPATHIC INTRACRANIAL HYPERTENSION: THE UNWANTED RISE

Idiopathic intracranial hypertension (IIH) is a syndrome caused by raised intracranial pressure (ICP) in the absence of any identifiable intracranial pathology as supported by normal neurological imaging. Previously, several terminologies were used to describe this condition. The most well-known but potentially misleading terms were pseudotumour cerebri and benign intracranial hypertension.[1,2] The latter is no longer used due to the awareness of potentially severe visual impairment. IIH commonly occurs in young women with a high body mass index. It has also been reported to occur in males and prepubescent age groups.[3,4]

The exact pathophysiology is unknown, but IIH is associated with several risk factors including obesity, obstructive sleep apnea (OSA), kidney failure, systemic lupus erythematosus and Behcet's disease. Several medication linked to the development of IIH includes tetracycline, oral contraceptives, cyclosporine, lithium, nalidixic acid, nitrofurantoin, danaxol, vitamin A and tamoxifen. Initiation and discontinuation of corticosteroids also has been indicated as a risk factor.[5,6]

Prior art suggests several pathogenesis which include imbalance between cerebrospinal fluid (CSF) absorption and production, elevation of cerebral venous pressure and high water composition of the brain.[7] Obesity seems to be a strong association of IIH. It has also been suggested that the accumulation of adipose tissue causes increased intra-abdominal and intra-thoracic pressure leading to raised central venous pressure and subsequently, high ICP.[8]

The typical clinical features of IIH are chronic headache with papilloedema with or without visual impairment. It has been previously reported that the incidence of debilitating or bilateral loss of vision due to IIH can be up to 10%.[9] Patients with papilloedema can have several visual field defect patterns. The commonest one is an enlarged blind spot from compression or disturbance of the peripapillary retina. Other patterns of field loss that have been described are peripheral constriction, arcuate scotomas and global reduction of sensitivity.[10]

IIH also may lead to visual impairment due to ocular changes other than papilloedema. Choroidal folds and subretinal fluid may form around the nerve head in fulminant IIH. Patients may also experience a hyperopic shift due to the flattening effect of the globe by the distended retrobulbar optic nerve sheath. Diplopia might also be present as a false localizing sign.[11] Other common visual complaints are transient obscurations of vision (TOV) where patients experience intermittent clouding of vision which lasts a few seconds. TOV is more prominent when changing position from supine to upright. This occurs due to momentary ischemia of the optic nerve axons.[12]

Headaches in IIH occur due to stretching of the meninges secondary to raised ICP. The headache is non-specific, however it is usually severe and may wake the patient up from sleep. Patients report having a headache which waxes and wanes and is exacerbated with valsalva manoeuvres.[13]

The best modality to view changes in IIH is MRI of the brain and vessels. A good image can show the flattened appearance of the posterior globe due to pressure from the swollen and tortuous optic nerve sheath. Severe elevation of ICP also may cause empty sella turcica.[7] On plain CT scanning, the appearance of slit-like / narrowed ventricles is a feature.

According to a Cochrane review in 2015, there are still no accepted guidelines in the management of IIH.[14] The objective of treatment is to preserve visual function and prevent blindness and secondarily to lessen the debilitating chronic headache experienced by patients.

Most of the cases of IIH are usually mild without visual impairment. A single lumbar puncture may dramatically improve the symptoms of headache.

Management of the modifiable risk factor are very important in the long term treatment i.e. treating the obesity and OSA. Discontinuation of the associated drugs such as tetracycline, and oral contraceptive pills will help with the remission.[11]

Common systemic medication given for the treatment of IIH include carbonic anhydrase inhibitors like acetazolamide. Its mode of action is to reduce CSF production thus reducing ICP. The idiopathic IHH treatment trial (IIHTT) showed that acetazolamide (4g/day) with a low sodium weight-reduction diet showed considerable improvement in visual function and improvement in the grading of papilloedema.[15] Topiramate is another drug of choice for its dual effect of being a carbonic anhydrase inhibitor and appetite suppressant for weight management.[16]

Surgical intervention is only indicated in patients with refractory IIH suffering from intractable headaches and advanced and progressive blindness, who have failed to respond to pharmacological treatment. Optic nerve sheath decompression is a method which involves optic nerve sheath incisions to release CSF into the retro-orbital space.[12] A meta-analysis regarding optic nerve sheath decompression done on more than 700 patients showed promising results with improvement in vision, resolution of headache, and reduction in papilloedema.[17] Other surgical methods including ventriculo-peritoneal or lumbar-peritoneal shunts and transverse venous stenting have not shown similar efficacy.[12]

Conclusion:

IIH is an uncommon disease. Although it is benign in most cases, it has the potential of causing debilitating visual loss and significant headache. Detection and management of modifiable risk factors remains the mainstay of treatment of IIH.

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