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Systemic review idiopathic intracranial Hypertension

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Abstract
Idiopathic intracranial hypertension is a rare disorder of intracranial hypertension with normal cerebrospinal fluid (CSF) and no pathology. The research aims to describe the disease, the factors affecting it, and treatment methods. To achieve the desired goal, we followed the descriptive approach by reviewing the scientific research published in this regard. The study concluded that headache is one of the most common symptoms, obesity is the main factor in its occurrence, and the best method of treatment is the use of medication along with weight loss.

Keywords: Idiopathic intracranial, CSF, Hypertension, IIH

Introduction

Idiopathic intracranial hypertension (IIH) is an uncommon neurological disorder of elevated cerebrospinal fluid (CSF) pressure in the absence of any intracranial pathology or secondary causes [1]. The incidence of IIH increases with obesity, especially in women of childbearing age [2]. The main symptoms include headache, vision problems, ringing in the ears synchronous to the pulse, and shoulder pain. Complications may include loss of vision [3]. Diagnosis is based on symptoms and high intracranial pressure observed during a lumbar puncture without a specific cause on a brain scan [1]. Treatment includes a healthy diet, salt reduction, and exercise. In 1893, Quincke was the first to describe a group of patients with symptoms and signs of raised intracranial pressure (ICP) without any cerebrospinal fluid abnormalities, which he termed "meningitis serosa" [3]. Later he termed pseudotumor cerebri. Benign hypertension existed until 1989 when Corbett and Thompson termed it "idiopathic intracranial hypertension." [3]. In this review, we will discuss the brief history of IIH, details about the pathophysiology, and prognosis of IIH.

Pathophysiology of idiopathic intracranial hypertension

The risk of developing IIH increases in people who suffer from weight gain, whether it is a slight increase of 5-15% or whether the body mass index is high [5]. This is explained by the following theory:

- **Increased central venous pressure:** Central obesity leads to elevated intra-abdominal, pleura, cardiac filling, and central venous pressure, which impairs cerebrospinal fluid flows [6].
- **Metabolism:** Insulin resistance and hyperlipidemia in centripetal obesity are associated with increased opening pressure at lumbar puncture, yet it remains unknown [7].
- **Hormonal changes:** Neuroendocrine adipose tissue releases cytokines and leptin [5].

High leptin in the blood leads to a defect in the regulation of the intracellular enzyme (11β-HSD1), which affects cerebrospinal fluid production and homeostasis through its expression in the epithelium of the choroid plexus and arachnoid granulation tissue [8]. Narrowing of the dural venous sinus impairs venous outflow, leads to venous hypertension, and increases intracranial hypertension. Figure (1) shows the pathophysiology of idiopathic intracranial hypertension.

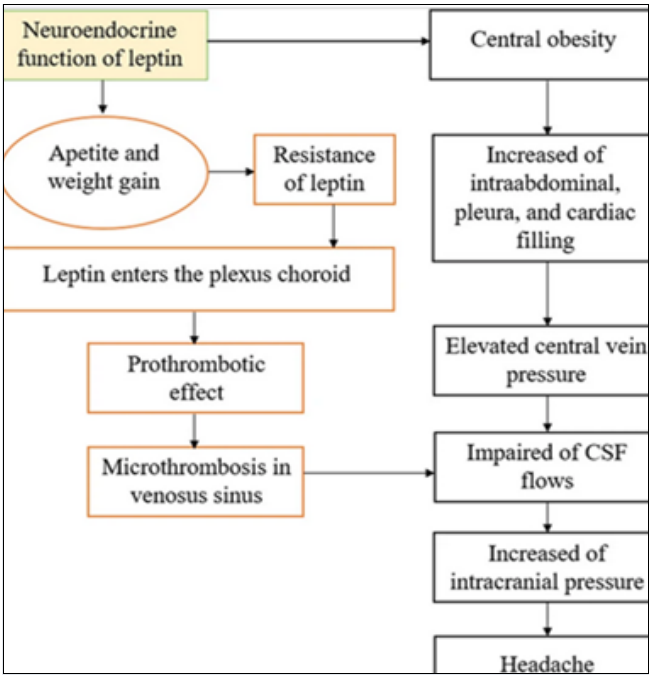

Fig 1: pathophysiology of idiopathic intracranial hypertension (source: [3])

Factors related to idiopathic intracranial hypertension

The underlying causes of IIH are unknown. However, some factors play a role in the patient becoming infected with it. Important risk factors that have been identified are

- Signs and symptoms resulting from weight gain [9].
- Medicines such as vitamin A derivatives and tetracycline [3].
- Endocrine diseases (thyroid dysfunction, Addison's disease, or hypoparathyroidism), levothyroxine, recombinant growth hormone, and anabolic steroids. Women can suffer from IIH in the first trimester of pregnancy [10].
- It is considered one of the associated complications of spine surgery [11].
- Anemia, because iron is a regulator of thrombopoiesis, anemia, specifically iron deficiency anemia, results in elevated platelet levels and a hypercoagulable state. In addition, iron shortage causes abnormalities in red blood cells. [12].

Clinical manifestation

- Frontal and periorbital pain: increased by eye movement brought on by intracranial hypertension-induced distension of the optic nerve sheath [13].
- Headache is the most common symptom, felt gradually in the morning or while lying down. There are different types of headaches, such as throbbing headaches, trigeminal neuralgia, referred pain, or migraine [14].
- Temporary vision impairment resulting from axoplasmic flow disruption, which causes retinal nerve fiber edema and short-term optic nerve ischemia 1 [15]
- Causes of pulsatile tinnitus include higher CSF flows in the cochlea, high standard vascular pulse transmission, and turbulence of blood flows in occluded transverse veins. It can manifest unilaterally or bilaterally [15].

Procedures of diagnostic

- A neurological examination of visual functions is performed, as papilledema is a feature of this disease [16]. Papilloedema may be difficult to identify using conventional noninvasive direct ophthalmoscopy because it requires significant ability and practice to visualize the optic disc [3]. Figure 2 Shows levels of Papilloedema.
- Dyspigmentation is an early sign of optic nerve dysfunction and can be detected using the Ishihara test. The patient usually does not complain of changes in the visual field and they are not noticeable to the visual field. Because it is an important clinical sign, it should be examined using an automated contour with a fixed threshold to measure the mean deviation (MD; typical < -2 decibels or dB) [5].
- In addition, extraocular movements should be checked for abductor palsy, which indicates increased intracranial pressure [6].
- characteristic radiological changes that can provide important clinical information, such as the elevation of the optic nerve surrounding the cerebrospinal fluid, empty sella turcica, and narrowing of the transverse sinuses [17].

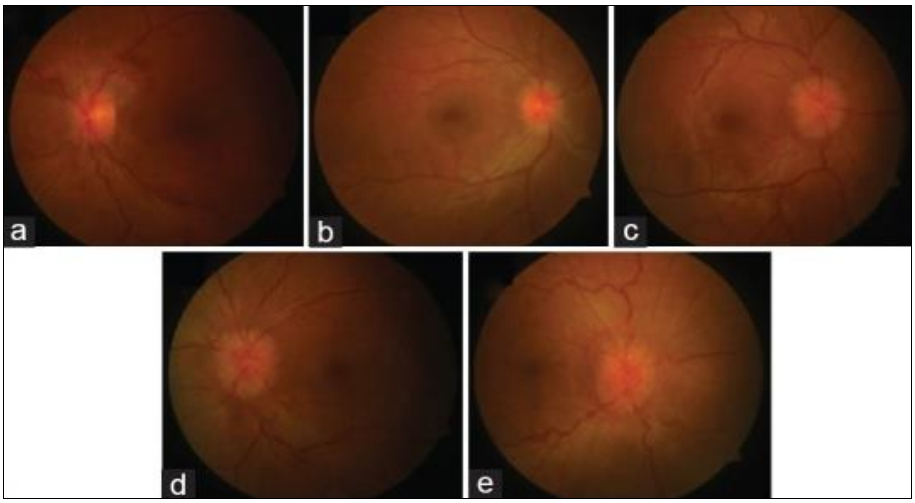

Fig 2: levels of Papilloedema. (source: [4])

Treatment of idiopathic intracranial hypertension

To treat IIH, any risk factors such as obesity, drug abuse, and endocrine diseases must be identified. Treatment includes body weight control, and medical and surgical therapy depending on severity, visual function, comorbidity, response, and tolerability to treatment [18].

- **weight control:** Conservative treatment Patients with a high body mass index should follow a diet to reduce weight by 6-15% over 6 months. In the event of failure to lose weight through diet and exercise, surgery may be necessary, as weight loss can reduce papilloedema or treat it [5].
- **The medical treatment:** It is recommended to use acetazolamide, which plays a role in reducing the secretion of cerebrospinal fluid in the choroid plexus. Trials have shown that the use of acetazolamide in combination with weight loss leads to improvement in papilledema and visual function compared with IIH patients who had weight control without medical treatment [19]. The glucagon-like peptide-1 (GLP-1) agonist, exendin-4, also reduces intracranial pressure. Moreover, the 11β-HSD1 inhibitor (AZD4017) can change from inactive cortisol to active cortisol, thus regulating blood cortisol and affecting cerebrospinal fluid production [14]

- **Surgical treatment:** *Surgical* treatment is resorted to in some cases that include gradual visual impairment or severe vision loss, failure to respond to medical treatment, or prolonged symptoms (refractory headache). Surgical treatment options include:

a. Sinus venous stent: The first-line surgical procedure for treating refractory IIH is venous sinus stenting. About 10% of the procedures were repeated with only a minor 1.5% serious complication rate, and this intervention had a high technical success rate of about 99.5%. With minimal failure and advantageous consequences, this method produced the best clinical results in terms of headache and visual function [20].

b. CSF diversion methods: Cerebrospinal fluid shunts for ocular preservation can be carried out by decreasing the volume of the lumboperitonial and ventriculoperitoneal shunts cerebrospinal fluid [21]. With a minimum complication rate of roughly 9.4% and a failure rate of 43.4%, the cerebrospinal fluid diversion approach improved papilledema, visual field, and headache in 78.9%, 66.8%, and 69.8% of cases [20].

c. Optic nerve sheath fenestration: ONSF lessens the damage that increased intracranial pressure causes to the optic nerve by creating a fistula between the optic nerve sheath. In certain circumstances, such as a mild headache, high risk, or when CSF diversion techniques are contraindicated, it is suggested [3]. With a minimal severe complication rate of 0.2% and a failure rate of 9.4%, papilledema, visual field defects, and headaches can improve in 90.5, 65.2%, and 49.3% of cases, respectively [20].

d. Bariatric surgery: Bariatric surgery was also excellent for reducing intracranial pressure compared to weight management in a randomized clinical trial conducted by Mullan and colleagues in 2021. It also improved other symptoms, such as headache and vision [22].

Conclusion

Intracranial hypertension is accompanied by symptoms such as headache and gradual loss of vision. There is no clear cause for the patient's injury, but obesity is considered one of the main causes. There are different treatment methods, including medication (acetazolamide, peptide-1, exendin-4), surgical (Sinus venous stent, Bariatric surgery, ONSF, Cerebrospinal fluid shunts), and weight loss.

Reference

1. Dhungana S, Sharrack B, Woodroofe N. sIdiopathic intracranial hypertension," *Acta Neurologica Scandinavica*,2010:121(2):71–82. doi: https://doi.org/10.1111/j.1600-0404.2009.01172.x.
2. Boyter E. Idiopathic intracranial hypertension," *Journal of the American Academy of Physician Assistants*,2019:32(5):30–35. doi: https://doi.org/10.1097/01.jaa.0000554732.85914.91.
3. Made I, Tiara C. Idiopathic intracranial hypertension: from concise history to current management," *The Egyptian Journal of Neurology, Psychiatry and Neurosurgery*, 2023, 59(1). doi: https://doi.org/10.1186/s41983-023-00730-7.
4. Lal V, Takkar A. Idiopathic intracranial hypertension: The monster within," Annals of Indian Academy of *Neurology*, 2019. doi: https://doi.org/10.4103/aian.aian_190_19.
5. Thurtell Matthew J, Aki Kawasaki. Update in the Management of Idiopathic Intracranial Hypertension." *Neurologic Clinics*,2021:39(1):147–161, https://doi.org/10.1016/j.ncl.2020.09.008.
6. Wakerley, Benjamin R, *et al*. Idiopathic Intracranial Hypertension: Update on Diagnosis and Management." *Clinical Medicine*,2020:20(4):384–388. www.ncbi.nlm.nih.gov/pmc/articles/PMC7385768/, https://doi.org/10.7861/clinmed.2020-0232.
7. Westgate Connar SJ, *et al*. Systemic and Adipocyte Transcriptional and Metabolic Dysregulation in Idiopathic Intracranial Hypertension." *JCI Insight*, 6(10), e145346, www.ncbi.nlm.nih.gov/pmc/articles/PMC8262372/, https://doi.org/10.1172/jci.insight.145346.
8. Rim Rakez, *et al*. The Diagnostic Dilemma of Idiopathic Intracranial Hypertension in a Child with Acute Lymphoblastic Leukemia: COVID-19 or Cytosine Arabinoside?" *BMC Neurology*, 2022, 22(1). https://doi.org/10.1186/s12883-022-02689-z.
9. Zafar Sana, *et al*. A Systematic Review on Whether an Association Exists between Adolescent Obesity and Idiopathic Intracranial Hypertension." Cureus, 2022. https://doi.org/10.7759/cureus.28071.
10. Negro A, *et al*. "Headache and Pregnancy: A Systematic Review." The Journal of Headache and Pain, 2017, 18(1). www.ncbi.nlm.nih.gov/pmc/articles/PMC5648730/, https://doi.org/10.1186/s10194-017-0816-0.
11. Xu Zhengkuan, *et al*. Idiopathic Intracranial Hypertension Occurred after Spinal Surgery: Report of Two Rare Cases and Systematic Review of the Literature, 2016. https://doi.org/10.1007/s00586-016-4578-1.
12. Ma Zhonghua, *et al*. Idiopathic Intracranial Hypertension in Patients with Anemia: A Retrospective Observational Study." PLOS ONE,2020:15(7):e0236828, https://doi.org/10.1371/journal.pone.0236828.
13. Friedman Deborah I. Headaches in Idiopathic Intracranial Hypertension." *Journal of Neuro-Ophthalmology*,2019:39(1):82–93, https://doi.org/10.1097/wno.0000000000000777.
14. Hasırcı Bayır, Buse Rahime, *et al*. Current Overview of Idiopathic Intracranial Hypertension." *Turkish Journal of Neurology*,2021:27(3):219–228, https://doi.org/10.4274/tnd.2021.45649.
15. Lal Vivek, Aastha Takkar. Idiopathic Intracranial Hypertension: The Monster Within." *Annals of Indian Academy of Neurology*, 2019. https://doi.org/10.4103/aian.aian_190_19.
16. Zainab Rissan, *et al*. Fundus Imaging and Perimetry in Patients with Idiopathic Intracranial Hypertension—an Intermethod and Interrater Validity Study." *European Journal of Neurology*,2023:30(7):1973–1982. https://doi.org/10.1111/ene.15802.
17. Moreno Mauricio E, Raquel Del. Neuroimaging in the Diagnosis and Treatment of Intracranial Pressure Disorders." *Neurological Sciences*,2022:44(3):845–858. https://doi.org/10.1007/s10072-022-06478-x.

18. Biousse Valérie, *et al*. Update on the Pathophysiology and Management of Idiopathic Intracranial Hypertension." *Journal of Neurology, Neurosurgery & Psychiatry*,2012:83(5):488–494, https://doi.org/10.1136/jnnp-2011-302029
19. Wall Michael, *et al*. Effect of Acetazolamide on Visual Function in Patients with Idiopathic Intracranial Hypertension and Mild Visual Loss." *JAMA*,2014:311(16):1641. https://doi.org/10.1001/jama.2014.3312.
20. Kalyvas, Aristotelis, *et al*. A Systematic Review of Surgical Treatments of Idiopathic Intracranial Hypertension (IIH)." *Neurosurgical Review*,2020:44(2):773–792, https://doi.org/10.1007/s10143-020-01288-1.
21. Tuță Sorin. Cerebral Venous Outflow Implications in Idiopathic Intracranial Hypertension—from Physiopathology to Treatment." *Life*,2022:12(6):854, https://doi.org/10.3390/life12060854.
22. Sun Warren YL, *et al*. Idiopathic Intracranial Hypertension and Bariatric Surgery: A Systematic Review." *Canadian Journal of Surgery*,2020:63(2):E123–E128, https://doi.org/10.1503/cjs.016616.