

Non-idiopathic-intracranial hypertension: A case report

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ABSTRACT

Idiopathic intracranial hypertension (IIH) is a condition characterized by headache, visual obscuration, and pulsatile tinnitus caused due to raised intracranial pressures without any underlying pathology of brain parenchyma or its structure. The diagnosis of IIH can be reconsidered especially when there are no risk factors associated during presentation. This is a case report of a 27-year-old young girl with normal body mass index (BMI), who presented with persistent headache and tinnitus. Fundus examination revealed papilledema, with magnetic resonance imaging of the brain and orbit showing flattening of the posterior sclera and tortuous optic nerve. A patient with a normal BMI should always raise suspicion for secondary causes of IIH. She was using topical retinol gel for acne which is available easily over the counter. It is therefore important to raise awareness for such agents. Recognizing and discontinuation of these offending agents can prevent the progression and reversal of IIH symptoms.

Key words: Headache, Idiopathic intracranial hypertension, Tinnitus

Pseudotumor cerebri or idiopathic intracranial hypertension (IIH) is a condition characterized by headache, visual obscuration, and pulsatile tinnitus caused due to raised intracranial pressures (ICP) without any underlying pathology of the brain parenchyma or its structure. The patient shows no signs of altered sensorium or level of consciousness. Complications of this disease include blindness, and therefore, the initial term benign intracranial hypertension was renamed as IIH due to its not-so-benign nature [1]. IIH was reported in children as young as 4 months, with no gender predisposition. Children with IIH have poor performance in school. Prospective studies show an incidence of 1–2/100,000 females>males), especially in females with the childbearing age group. High body mass index (BMI >25), obesity related to increased intra-abdominal adipose tissue, polycystic ovary disease in females, and androgen deficiency in males are the risk factors associated with IIH. There is a positive correlation between elevated BMI and the risk of IIH, irrespective of gender [1]. Vitamin A derivatives, tetracycline-class antibiotics, recombinant growth hormone, and lithium (Categories IV and V) and corticosteroids (Category III) contribute to drug-induced IIH [2,3].

CASE PRESENTATION

A 27-year-old young Asian female presented in the outpatient department with severe headache and blurring of vision

progressing for 3 days without any fever, vomiting, or altered behavior. She had a frontal headache with periorbital pain, pulsatile tinnitus, and neck pain. Her pain did not get relieved with over-the-counter medications.


On examination, BMI was 22.6 kg/m², and grade 3 papilledema was seen with no loss of color vision, anisocoria, visual field defect, and signs of meningeal irritation.

Her routine blood investigations and hormonal evaluation including cortisol levels, luteinizing hormone, follicle-stimulating hormone, prolactin, and progesterone were normal.

Magnetic resonance imaging (MRI) brain with orbit (contrast) showed prominent cerebrospinal fluid (CSF) perioptic space around bilateral optic nerves, prominent Meckel's cave, attenuated dural venous sinuses in the lateral part of bilateral transverse sinuses, tortuous optic nerves, and flattened posterior sclera (Figs. 1 and 2). Ophthalmology evaluation revealed decreased visual acuity and bilateral disc edema.

She was taken for lumbar puncture (LP) and the pressure recorded by manometer was 69 cm of H₂O. After draining 20 cc of CSF, there was a slight subjective improvement in headache and vision. CSF examination findings were normal for infection/inflammation. She was started conservatively with acetazolamide, mannitol, and furosemide. Three days post-LP, her vision improved with fewer episodes of headache.

She was discharged on the 5th day of admission, on acetazolamide with marked improvement in vision without headache. On follow-up, after 7 days, she revealed to have been

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Figure 1: Tortuous optic nerves

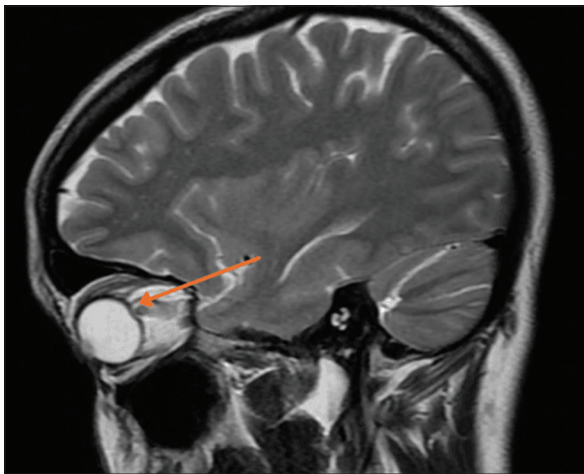


Figure 2: Flattening of posterior sclera

using transdermal formulation for acne, retinol gel (retinoic acid concentration 0.5%) for 2–3 months.

DISCUSSION

The incidence and prevalence of IIH are increasing with an increase in obesity and lifestyle changes. It has a predilection in the female population, especially with abdominal obesity and in the reproductive age group [1]. IIH can present with headaches, mostly early morning headaches, and increases on physical straining maneuvers such as coughing, passing stools, and lying down position. Visual abnormalities can be presented as blurring of vision, transient visual disturbances, and double vision due to the involvement of the abducens nerve. This can cause sleep disturbances, cognitive impairment which can mislead the actual diagnosis. Olfactory dysfunction, tinnitus, can be a rare manifestation of IIH [2,3].

The current understanding of IIH is limited. It is, however, presumed that dysregulation of CSF flow, secreted by choroid plexus, intracranially is the possible cause. It could be either an increase in its production or a reduction in its absorption which causes an increase in central venous pressure. The involvement of metabolic and hormonal imbalance, especially

excessive androgen, is related to CSF dysregulation. This has been identified lately giving the site of target for novel therapies and drug invention. The treatment aims to alleviate and prevent vision loss. The medical management includes acetazolamide with weight loss as the primary objective [4-6]. Therapeutic LP is done for the management of headache associated with IIH. It is presumed to alleviate headache but 64% of the patients have exacerbation of headache following LP. Fulminant or medically refractory patients are ideal for surgical intervention such as optic nerve fenestration, CSF diversion, or venous shunting [7].

Modified Dandy diagnostic criteria are followed for diagnosing IIH which comprises CSF pressure >25 cm of H₂O, normal CSF composition, signs and symptoms of raised ICP like papilledema, no focal deficits, and neuroimaging (empty sella, tortuous optic nerve, and flattened posterior sclera) with normal ventricular system structure and physiology [3]. The study done in the Danish and Austrian populations showed that MRI findings—flattening of the posterior globe, protrusion of optic nerve disc, and transverse sinus stenosis, can be a better diagnostic marker than the present diagnostic criteria, which emphasize the raised ICP [8].

The literature relating the transdermal application of retinol as the cause of IIH is not documented. Abnormal Vitamin A metabolism is hypothesized to increase the levels of retinol in CSF by reducing retinol-binding protein [9]. Our patient was having normal BMI, known to be the primary risk factor in IIH. This case report emphasizes the use of topical Vitamin A as a possible cause of IIH especially when there are no other risk factors (increased BMI, normal morning cortisol levels) associated.

CONCLUSION

IIH does not always get along with its nomenclature as idiopathic. There could be hidden triggers which the patient might feel unrelated to the disease or can presume to be insignificant. A patient with a normal BMI should always raise suspicion for secondary causes of IIH. Therefore, it is important to raise awareness for offending agents. If these can be recognized, discontinuation of these agents can prevent the progression and reversal of symptoms.

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