

Idiopathic Intracranial hypertension: A case report and a brief review of literature

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Abstract: We report a case of Idiopathic intracranial hypertension and Obesity. An obese female presented with headache and diplopia, since 2 weeks. She recovered very well with diamox and short steroid therapy. IIH is often idiopathic, proposed to be due to impaired cerebrospinal fluid absorption from the sub-arachnoid space across the arachnoid villi into the dural sinus. IIH is common in obese females and can lead to visual impairment but prompt diagnosis and treatment in most of the cases will prevent potentially permanent visual loss.

Key Words: Idiopathic intracranial hypertension, Obesity.

I. Introduction

Idiopathic intracranial hypertension (IIH) is a headache syndrome characterized by raised cerebrospinal fluid pressure in the absence of any cranial mass lesion or ventricular dilatation, normal spinal fluid composition, usually normal findings on neurological examination except for papilloedema and an occasional VI nerve palsy, and normal level of consciousness. IIH is common in obese females and can lead to visual impairment. Prompt diagnosis and treatment are needed to prevent potentially permanent visual loss. The incidence of IIH in obese women was 11.9 per 1,00,000 per year. Upto 90 % patients were overweight.

II. Case Report

A 50 years old female presented with global headache and diplopia on the left side for which she went to an ophthalmologist. She was given treatment for 8 days but there was no relief and she came to us with chief complaints of headache and diplopia since 10 days.

On clinical examination, the patient was obese with weight of 121kg and height of 165 cm (BMI of 44). Patient was conscious, oriented, had a pulse rate of 80 / min and blood pressure of 130/80 mmHg. She had horizontal diplopia on left gaze. Central nervous system examination was normal. Patient was diagnosed case of Diabetes Mellitus type II, Hypertension and Ischemic Heart disease since last 10 years and on regular treatment with anti-diabetic, anti-hypertensive and aspirin therapy. She had no history of consumption of oral pills or any other medicine in the past. On admission, the blood pressure and blood sugar values were normal. Investigation revealed HbA1c of 6.7. Thyroid functions were normal. Ultrasound of abdomen showed fatty liver. Fundus examination showed bilateral disc edema (papilloedema). Perimetry showed borderline normal study. MRI brain was not suggestive of ventricular dilatation and there was hypoplastic sigmoid sinus on the left side. MR angiography and MR venography were also normal.

Clinical suspicion of Idiopathic intracranial hypertension was confirmed. She was put on Diamox and short steroid therapy, she improved.

Patient made a good recovery after Diamox and steroid therapy at one month.

On discharge she was advised a 35 mins walk per day 5 times a week. Anti-obesity measures in the form of voglibose, exercise and diet control were prescribed which reduced her weight in 2 months from 121 kg to 113 kg.

Patient has been under follow up and after 1 month of treatment, she did not have any complaints of

diplopia and headache. This case has been presented with a view to highlight the entity of Idiopathic intracranial hypertension in an obese female.

III. Discussion

We have discussed a case of idiopathic intracranial hypertension with obesity. IHH is a syndrome characterized by elevated intracranial pressure that usually occurs in obese women of childbearing age¹. Foley coined the term Idiopathic intracranial hypertension in 1955 but reports from the 1980's demonstrated the high incidence of visual loss^{2,3}. The annual incidence of BIH is 0.9/100000 person and 3.5/100000 in females 15-44 years of age. It is increasing in incidence in parallel with the current epidemic of obesity^{4,5}. Generally causes of IHH are endocrine⁶ like adrenal insufficiency, Cushing's syndrome, hypoparathyroidism, hypothyroidism and hyperthyroidism, drugs like doxycycline, levothyroxine, lithium, nitrofurantoin, tamoxifen, tetracycline, pancreatin and chronic renal failure and SLE⁷. A case control study has found a strong association between IHH and obesity and with weight gain during the 12 months before the IHH diagnosis. In this study there was no evidence the IHH was associated with any other medical condition or pregnancy⁸. There was no drug history in the past to suggest causation of IHH.

The symptoms of Idiopathic intracranial hypertension patients are headache (94%), transient visual obscurations (68%), pulse synchronous tinnitus (58%), photopsia (54%), and retrobulbar pain (44%). Visual loss and diplopia are less common with Idiopathic intracranial hypertension. Headache, nausea, vomiting and visual disturbances are the most common presenting features⁹. The headache profile of the IHH patient is that of severe daily pulsatile headache which is generally throbbing, worst in the morning hours and aggravated on straining, coughing, or changing of posture¹⁰.

Visual obscurations are episodes of transient blurred vision of 30 seconds and are followed by visual recovery to baseline. Visual obscurations occur in about 2/3rd of Idiopathic intracranial hypertension patients¹¹. Pulsatile intracranial noises or pulse synchronous tinnitus is common in Idiopathic intracranial hypertension. Papilloedema, optic disc edema due to increased intracranial pressure is the cardinal sign of Idiopathic intracranial hypertension. Optic disc edema either directly or indirectly is the cause of visual loss of Idiopathic intracranial hypertension. The higher the grade of papilloedema, the worse is the visual loss¹².

Horizontal diplopia is reported in about 1/3rd of Idiopathic intracranial hypertension patients and 6th nerve palsy are found in 10-20%¹³.

On perimetry, visual field loss occurs in almost all cases of Idiopathic intracranial hypertension in later stages. Visual loss in almost one eye was found in 96% of patients with Goldmann perimetry using a disease specific strategy and in 92% with automated perimetry¹⁴.

There is currently no consensus on the management of IHH¹⁵. Management is initially medical with weight reduction if obese and diuretic therapy. CSF diversion therapy may be required for visual disturbances¹⁶.

The aim of treatment is relief of all the symptoms of raised ICT and prevention of progression of optic nerve damage¹⁷. Since marked weight gain is a predictor of visual disorientation and papilloedema can resolve with modest weight loss, institution of low salt diet and regular exercise appears to be beneficial for Idiopathic intracranial hypertension patients.

Acetazolamide a carbonic anhydrase inhibitor is the drug of 1st choice. It has been shown to reduce papilloedema and decrease the CSF pressure^{18,19}. Mac Carty and Reed²⁰ showed that acetazolamide reduces the CSF flow. Steroids are still used to treat Idiopathic intracranial hypertension but their mechanism of action remains unclear. Surgical interventions may be used if the other methods are ineffective. Surgical options include²¹:

- * CSF diversion (lumbo-peritoneal or ventriculo-peritoneal shunt)
- * Intracranial venous sinus stenting
- * Optic nerve sheath fenestrations (decompression)

Optic nerve sheath fenestration is now the currently favoured treatment of Idiopathic intracranial hypertension in adults with deteriorating visual functions despite medical management. The procedure successfully relieves papilloedema, rapidly recovering visual loss in most cases^{22,23}.

A study of EJ Rowe²⁴ and NJ Sarkies²⁵, to evaluate the association between obesity and IHH whether there is a relationship between the visual outcome, 70.5% patients were found. A high degree of obesity was associated with poor visual outcome and identified as a risk factor for poor outcome. A prospective study by Wall and George²⁶ showed recent weight gain was significantly associated with worsening vision. Ireland et al²⁷ have reported a significant weight gain in IHH patients during the 6 months preceding diagnosis and their study confirmed a strong association between IHH and obesity and weight gain during the 12 months before the diagnosis.

IV. Conclusion

Idiopathic intracranial hypertension is characterized by elevated CSF pressure of unknown cause. It is predominantly a disease of women in the childbearing age. Although the cause of IHH remains obscure, it has become clear that the loss of visual functions is common and the patients may progress to blindness if untreated. Most of the cases respond to acetazolamide and short course of steroids coupled with weight reduction by exercise and diet control.

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