

Case report

Idiopathic Intracranial Hypertension in a pubertal paediatric Indian patient

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Abstract

Background: Idiopathic Intracranial Hypertension (IIH) is characterised by raised intracranial pressure (ICP) with normal cerebrospinal fluid (CSF) composition and absence of hydrocephalus or space occupying lesions. IIH is a rare disease in children. It can lead to visual impairment but prompt diagnosis and treatment in most of the cases will prevent potentially permanent visual loss. **Objective:** To report a rare case of Idiopathic Intracranial Hypertension in a pubertal child, clinical features, and findings of Magnetic Resonance Imaging (MRI) and visual field of this case. **Case:** An adolescent girl aged 14 years presented with headache and transient visual obscuration for two weeks. On examination, findings (fundus, visual field and MRI) were suggestive of Idiopathic intracranial hypertension. She did not have any classical predisposing risk factors. She recovered very well with acetazolamide and short term steroid therapy with no sequelae and clinical recurrence over a follow up of 12 months. **Conclusion:** This is a rare case of IIH in a child, which was confirmed on the MRI and visual field testing.

Keywords: Idiopathic Intracranial Hypertension; Papilloedema

Introduction

IIH is a headache syndrome characterized by raised cerebrospinal fluid pressure in the absence of any intra cranial lesion or other underlying systemic cause (Friedman DI et al, 2002). It most frequently occurs in obese females of childbearing age but can occur in all age groups, both genders and both obese and non-obese individuals. The condition is infrequent in children.

Case Report

A 14 year old girl presented with global headache and transient visual obscurations for two weeks. Her headache was severe in intensity, throbbing in nature and was aggravated by exercise. It was progressive with increasing frequency up to several times a day. Her headache was associated with nausea and a single episode of vomiting. She also reported ringing sound in her ears that was synchronous with her pulse. There was no history of any preceding head injury, viral illness or use of any medication. Her birth history and past health related events were otherwise unremarkable.

On physical examination, she weighed 69 kg and had height of 160 cm (Basal Metabolic Index, BMI of 26.95 kg/mm²). Patient was calm, conscious, oriented, with pulse rate of

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80 per min and blood pressure of 130/80 mm Hg. There were no focal neurological deficits. Cutaneous examination did not reveal presence of any neurofibromas or cafe au lait spots which would have pointed towards diagnosis of Neurofibromatosis type 1.

Local ophthalmic examination showed a visual acuity of 20/20 in both eyes. Extraocular motility was full, without diplopia and extra ocular muscle limitation. Pupils were normal sized, reactive to light; with no relative afferent pupillary defect. Iris was normal coloured with normal pattern without any Lisch nodules. Intraocular pressure was 24 mm Hg in right eye and 21 mm Hg in the left. Colour vision was normal with Ishihara's pseudoiso-chromatic plates in each eye. External and slit lamp examination was normal. Fundus examination showed bilateral papilloedema (Grade 3). (Figure 1)

An automated perimetry was performed, which showed a mean deviation of -3.78 dB in right eye with enlarged blind spot. In the left eye, mean deviation was -3.00 dB with enlargement of the blind spot. (Figure 2)

A non-contrast T2 weighted MRI scan was performed which showed thickening of bilateral optic nerve with tortuosity of its sheath on the left side, slight bulge of the bilateral optic disc and normal sized ventricles. (Figure 3)

Lumbar puncture (LP) was done which revealed an opening pressure of 24 mm Hg with a normal CSF composition. Complete haemogram, serum cortisol, serum calcium and thyroid functions were all normal.

She was diagnosed as a case of idiopathic intracranial hypertension according to the Modified Dandy's Criteria (Smith JL, 1985) which includes symptoms and signs of raised intracranial pressure, absence of localising

neurological signs, raised cerebrospinal fluid pressure with normal CSF composition and normal sized ventricles.

The patient was recommended a low salt, weight reduction diet and exercise and was put on prednisolone 60 mg once a day and acetazolamide 500 mg twice a day for 1 week which was stepped up to 500 gram thrice a day. After 2 weeks of this treatment, patient showed significant improvement in her symptoms with partial resolution of papilloedema. Liver and renal function tests were normal. Corticosteroid therapy was gradually tapered and finally stopped at 1 month. The patient continued with acetazolamide for 6 months when she showed complete resolution of symptoms and signs. (Figure 4 and 5)



Figure 1: Fundus picture of the patient showing bilateral papilloedema

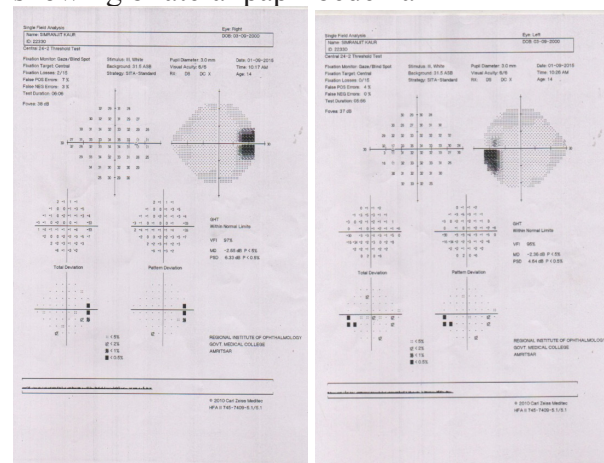


Figure 2: 24-2 Visual Field using SITA-standard strategy showing enlarged Blind spot in both eyes.

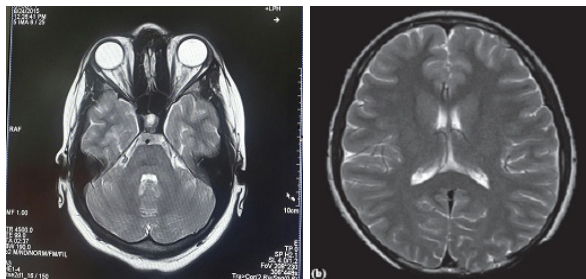


Figure 3. Non-contrast T2 weighted MRI showing thickening of optic nerves and normal sized ventricles.



Figure 4: Fundus picture showing resolution of papilloedema after 6 months of treatment.

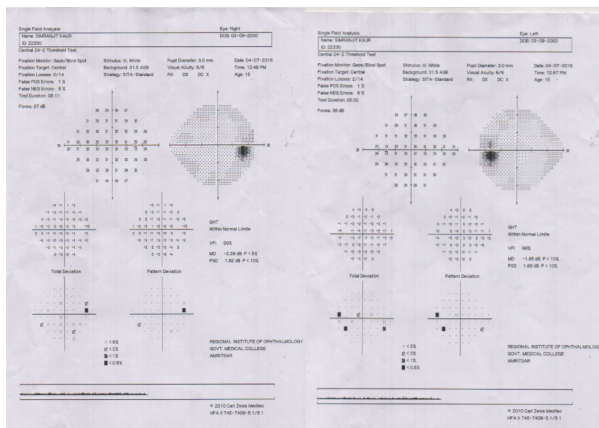


Figure 5. Visual field returning to normal after 6 months of treatment

Discussion

IIH is a rare disorder in childhood. Paediatric IIH is further divided into prepubertal and pubertal IIH based on the development of secondary sexual characteristics of age. In prepubertal IIH, there is no sex predilection or tendency toward obesity. In pubertal IIH, female predominance has been found. Association with obesity is also found in pubertal IIH (Wall M et al, 1991;

Babikian P et al, 1994; Kesler A et al, 2002; Phillips PH et al 1998).

The presenting symptoms of paediatric IIH are often non specific and may mimic migraine. Headache is typically generalised, throbbing in nature and aggravated on straining, coughing or change of posture (Wall M, 1990). It may be associated with nausea, vomiting, stiff neck, pulsatile tinnitus, retro-orbital pain, photophobia, dizziness or sensorineural hearing loss (Reitsma S et al, 2015). Neurological examination is normal with the exception of unilateral or bilateral sixth nerve palsies, which occur in 9-48% of children (Wolf A et al, 2008; Rangwala LM et al, 2007). Other cranial nerves including the 3rd, 4th, 7th, 9th and 12th have also been reported. Visual complications include transient visual obscurations, visual field defects and finally irreversible deterioration in visual acuity. High grade papilloedema, marked visual field loss, subretinal haemorrhage, hypertension and recent weight gain are risk factors for rapid progression and poor visual outcome (Spennato P et al, 2011; Standridge SM, 2010)

In children suspected of IIH, cranial imaging should be performed prior to LP to exclude a space occupying lesion in the brain. MRI is superior to computed tomography and the findings include tortuosity of the optic nerve, distension of perioptic subarachnoid space, posterior flattening of the globe or empty sella (Shin RK et al, 2001). CSF opening pressure of 25 mm Hg in children of 8 years or older, and opening pressure of 20 mm Hg in those younger than 8 years is diagnostic (Spennato P et al, 2011).

The aim of treatment is relief of all the symptoms of raised ICT and prevention of progression of optic nerve damage (Johnson LN et al, 1998). Acetazolamide, a carbonic anhydrase inhibitor, is the first line medication in treatment of IIH.

Recommended starting dose is 25 mg/kg/day given in three to four divided doses, with maximum dose of 100 mg/kg/day or 2 grams/day (Matthews YY, 2008). Steroids are used in the treatment of IIH but their mechanism of action is not clear. Therapeutic LP is used as an acute relief of symptomatic IIH but its effects are short-lived.

Majority of patients with IIH respond well to medical treatment. Most children have resolution of symptoms and papilloedema after one week to six months of treatment. Children who present with severe or rapid visual loss at onset, severe papilloedema with macular oedema and exudates, intractable headache and those who do not respond to medication with worsening of symptoms and signs are potential candidates for surgery (Wall M, 1990; Reitsma S et al, 2015). Surgical interventions include CSF diversion procedures such as lumboperitoneal or ventriculoperitoneal shunt, and optic nerve sheath fenestration (Reitsma S et al, 2015; Wolf A et al, 2008; Rangwala LM et al, 2007). Frequent follow-up, including visual field testing at intervals of 1 month, 3 months, and then every 3-6 months, is advisable (Passi N et al, 2013).

Our case of IIH presented with headache and transient visual obscurations with no focal neurological deficit. She showed significant improvement after 6 months of medical treatment with acetazolamide and short term oral corticosteroid with no sequelae and no clinical recurrence over a follow up of 12 months.

Conclusion

IIH is rare in children. They may present with non specific symptoms and the established risk factors are usually absent. Heightened physician awareness is important for early detection and prompt treatment to prevent irreversible visual loss.

References

- Babikian P, Corbett J, Bell W (1994). Idiopathic intracranial hypertension in children: the Iowa experience. *J Child Neurol*; 9(2):144-149.
- Friedman DI, Jacobson DM (2002). Diagnostic criteria for idiopathic intracranial hypertension. *Neurology*; 59(10):1492-1495.
- Johnson LN, Krohel GB, Madsen RW, March GA, Jr (1998). The role of weight loss and acetazolamide in the treatment of idiopathic intracranial hypertension (pseudotumor cerebri) *Ophthalmology*; 105(12):2313-2317.
- Kesler A, Fattal-Valevski A (2002). Idiopathic intracranial hypertension in the pediatric population. *J Child Neurol*; 17(10):745-748.
- Matthews YY (2008). Drugs used in childhood idiopathic or benign intracranial hypertension. *Arch Dis Child Educ Pract Ed*; 93(1):19-25.
- Passi N, Degnan AJ, Levy LM (2013). MR imaging of papilledema and visual pathways: Effects of increased intracranial pressure and pathophysiologic mechanisms. *Am J Neuroradiol*; 34(5):919-924.
- Phillips PH, Repka MX, Lambert SR (1998). Pseudotumor cerebri in children. *J AAPOS*; 2(1):33-38.
- Rangwala LM, Liu GT (2007). Pediatric idiopathic intracranial hypertension. *Surv Ophthalmol*; 52(6):597-617.
- Reitsma S, Stokroos R, Weber JW, van Tongeren J (2015). Pediatric idiopathic intracranial hypertension presenting with sensorineural hearing Loss. *Ann Otol Rhinol Laryngol*; 124(12):996-1001.
- Shin RK, Balcer LJ (2001). New developments in idiopathic intracranial



hypertension. *Curr Neurol Neurosci Rep*; 1(5):463-470.

Smith JL (1985). Whence Pseudotumor cerebri? *J Clin Neuroophthalmol*; 5(1):55-56.

Spennato P, Ruggiero C, Parlato RS et al (2011). Pseudotumor Cerebri. *Childs Nerv Syst* ;27(2):215-235.

Standridge SM (2010). Idiopathic intracranial hypertension in children: a review and algorithm. *Pediatr Neurol*; 43:377-390.

Wall M, George D (1991). Idiopathic intracranial hypertension. A prospective study of 50 patients. *Brain*; 114(Pt 1A):155–180.

Wall M (1990). The headache profile of idiopathic intracranial hypertension. *Cephalalgia*; 10(6):331-335.

Wolf A, Hutcheson KA (2008). Advances in evaluation and management of pediatric idiopathic intracranial hypertension. *Curr Opin Ophthalmol*; 19(5):391-397.

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