Original Research Paper Medical Science

Idiopathic Intracranial Hypertension: A Clinical Dilemma

DHARMANSHU CHAUBE	Resident,Department of Pediatrics,Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Dehradun, Uttarakhand
ALPA GUPTA	Professor, Department of Pediatrics, Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Dehradun, Uttarakhand
RENU DHASMANA	Professor, Department of Ophthalmology, Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Dehradun, Uttarakhand
MANJU SAINI	Professor, Department of Radiology, Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Dehradun, Uttarakhand

ABSTRACT

Idiopathic Intracranial hypertension (IIH) is a rare condition, incidence being 1 in 100,000 in general population and lesser in pediatric age group. Diagnosis requires high index of suspicion, and to rule out other causes of raised intracranial pressure. The treatment goal is to preserve optic nerve function while managing increased intracranial pressure. Optic nerve function should be carefully monitored with an assessment of visual acuity, color vision, field of vision and optic nerve head. Case characteristics: 13 year old adolescent girl weighing 45 kg presented in emergency with complaints of headache, fever and vomiting for 3 days. Fundus examination showed bilateral papilledema grade I. Opening pressure of CSF was raised but its cytological and biochemical assessment was normal. MRI brain showed prominence of subarachnoid space around bilateral optic nerve. The girl showed signs of rapid improvement with diuretics and was discharged. Resolution of papilledema was documented after 2 weeks.

KEYWORDS : Idiopathic intracranial hypertension; Pseudotumor cerebri

Introduction

Idiopathic Intracranial hypertension (IIH) is a rare condition. Its incidence in the general population is 1 of 100,000. Its prevalence among pediatrics age group is less known compared to the adults. Whenever present, it poses a clinical dilemma requiring intense clinical, investigative and multidisciplinary approach. Here we are presenting a case report of a young adolescent girl with Idiopathic Intracranial hypertension

Case report

A 13 year old, adolescent girl weighing 45 kg with normal BMI of 16.3kg/m^2 presented with complaints of headache, fever and vomiting. Her blood pressure and heart rate were normal. Headache was predominantly involving the frontal region. It was severe and incapacitating. There was no history of photophobia and blurring of vision. Vomiting was projectile, non-bilious and not associated with any other gastrointestinal symptoms.

There was no history of any drug, including Vitamin A, intake. There was no history of head trauma. General examination was markedly normal. Her ophthalmological examination revealed normal visual acuity, and normal color vision, her ocular movement were full and painless; and papillary reflex were normal but her fundus examination showed bilateral papilledema grade I. CNS examination had no evidence of any focal neurological deficit or any cranial nerve involvement. On lumbar puncture, the opening pressure was raised, but cytological and biochemical assessment of CSF was normal. MRI brain and orbit showed prominence of subarachnoid space around bilateral optic nerve with normal ventricular size [Fig.1&2]. There was no evidence of any space-occupying lesion of the brain. She was treated medically with oral Acetazolamide in a dose of 250 mg qid along with injection Mannitol (for initial 2 days). The girl showed rapid signs of improvement and was discharged when her headache and vomiting subsided. On follow up after 2 weeks she was asymptomatic and her fundus examination showed resolution of papilledema.

Discussion

Idiopathic Intracranial hypertension (IIH), was earlier known as Pseudotumor cerebri .It is characterized by signs and symptoms of increased intracranial pressure (ICP) in the absence of a space-occupying lesion.[1] In infants it is uncommon and there is an increasing incidence among adolescents as compared to young children[2]. One study reported that 60% of children who develop IIH are over 10 years of age[3]. It is more commonly seen in obese females of childbearing age[5] with strong predilection for postpubertal females. In children younger than 6 years, specific cause can be identified. Various causes commonly implicated are Hypovitaminosis A, Nitrofurantoin intake, Wiskott-Aldrich syndrome, Chronic renal insufficiency, Systemic lupus erythematosus, Congenital adrenal hyperplasia, Addison's disease, obesity, Turner syndrome etc. Idiopathic cases of intracranial hypertension are usually seen after the age of 11 years. It is a disorder defined by clinical criteria that include symptoms and signs isolated to those produced by increased intracranial pressure (e.g. headache, papilledema, vision loss). The intracranial pressure is considered increased when it is ≥ 280 mm Hg in sedated or obese children; ≥ 250 mm Hg in non-obese, non-sedated children. It is reported to have a normal cerebrospinal fluid (CSF) cell count and protein content. On MRI brain ventricular size may be normal to slightly decreased with normal ventricular anatomy and position. The pathophysiology of pseudotumor cerebri is not clearly understood but may include alterations in CSF absorption and production, subtle cerebral edema, abnormalities in vasomotor control and cerebral blood flow. Papilledema is the most consistent neurologic manifestation and is universally present. The goal of management for patients with idiopathic intracranial hypertension (IIH) is to preserve optic nerve function while managing increased intracranial pressure (ICP). Medical management is multipronged. Optic nerve function should be carefully and regularly monitored with an assessment of visual acuity, color vision, optic nerve head appearance and perimetry. Weight control is recommended for obese patients [5]. Acetazolamide appears to be the most effective agent for lowering ICP and most patients experience adequate relief of symptoms usally (headache) with this it. However the patients should be closely monitored while on medical treatment. Despite close monitoring and maximum medical treatment, some patients do experience deterioration of their visual function.

In this situation, surgical intervention may be required. Surgical intervention most often takes 1 of the following 2 general approaches: i) Optic nerve sheath fenestration (decompression), ii) Cerebrospinal fluid (CSF) diversion (like ventriculoperitoneal shunt). [6] A protracted course lasting months to years is common [7]. In most patients, symptoms worsen slowly. With treatment, there is usually gradual improvement and/or stabilization, but not necessarily recovery; many patients have persistent papilledema, elevated intracranial pressure (as documented on lumbar puncture), and residual visual field deficits. Relapse after treatment occurs in approximately 28% of patients [8].

Conclusion

Idiopathic intracranial hypertension is less frequent in prepubertal children than in adults or adolescents. Female predominance is noted. Early diagnosis and treatment is the key to management. If diagnosed early, visual acuity can be preserved with proper management. Even after initial relief, patient may require a long term follow up to prevent ocular manifestation from subtle relapse.

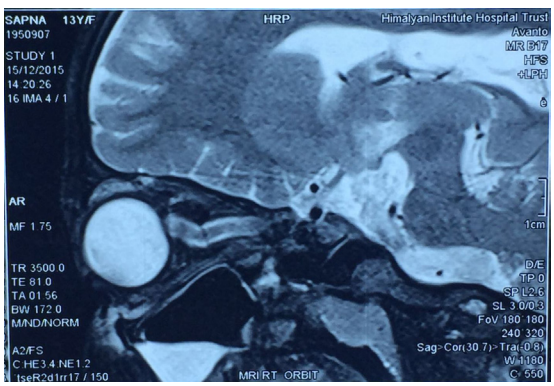

Figure 1: T2 weighted oblique sagittal section of right orbit showing prominence of subarachnoid space around optic nerve

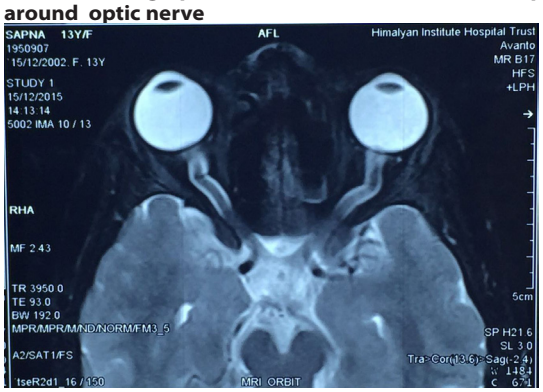

Figure 2: T2 weighted axial section of orbit showing prominence of subarachnoid space around bilateral optic nerve

References

1. Dessardo NS, Dessardo S, Sasso A, Sarunic AV, Dezulovic MS. Pediatric idiopathic intracranial hypertension: clinical and demographic features. Coll Antropol. 2010 Apr. 34 Suppl 2:217-21.
2. Gordon K: Pediatric pseudotumor cerebri: descriptive epidemiology. Can J Neurol Sci 1997;24:219–221.
3. Babikian P, Corbett J, Bell W: Idiopathic intracranial hypertension in children: the Iowa experience. J Child Neurol 1994;9:144–149
4. Jindal M, Hiam L, Raman A, Rejali D. Idiopathic intracranial hypertension in otolaryngology. Eur Arch Otorhinolaryngol. 2009 Jun. 266(6):803-6
5. Szewka AJ, Bruce BB, Newman NJ, Biousse V. Idiopathic Intracranial Hypertension: Relation Between Obesity and Visual Outcomes. J Neuroophthalmol. 2012 Jan 3
6. Mark S Gans, MD; Chief Editor: Andrew G Lee, MD. Idiopathic Intracranial Hypertension Treatment & Management; Medscape. 2016 Jan 28.
7. Wall M, George D; Idiopathic intracranial hypertension. A prospective study of 50 patients. Brain. 1991;114:155
8. Yri HM, Wegener M, Sander B, Jensen R. Idiopathic intracranial hypertension is not benign: a long-term outcome study. J Neurol 2012 5 886-894.