Case Report

Benign Intracranial Hypertension -Is it Really Benign Always?

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Abstract

Background: In children, benign or idiopathic intracranial hypertension (IIH) is uncommon. The presenting symptoms are usually variable, which attribute to delay in diagnosis. Frequently encountered symptoms include; headache, vomiting, visual impairment, neck pain and diplopia. IIH in pre-pubertal children has distinct characteristics from the adult form. Delayed diagnosis in children usually attributes to the inability in picking subtle visual changes, which can subsequently leads to permanent visual damage. **Objective:** We report cases of two adolescent girls having IIH who presented with papilloedema, could be managed with lumboperitoneal shunting which resulted in favorable outcomes in terms of improved visual acuity and relief of symptoms. **Case:** We report two cases of IIH, presented with headache and severe visual loss, which showed non responsiveness to medical management and improved with a lumboperitoneal shunt procedure. **Conclusion:** In IIH cases, enhanced awareness, prompt diagnosis and treatment are important, to avoid the risk of permanent visual damage. Urgent surgical intervention is essential in patients who do not respond to medical treatment.

Key words: Benign intracranial hypertension; papilloedema; lumboperitoneal shunt.

Introduction

Though pediatrics patients presenting to an emergency room with headaches are often attributed to common disorders, but rarely these headaches can be due to some serious underlying disease. Idiopathic intracranial hypertension (IIH) is a diagnosis of exclusion and characterised by raised intracranial pressure (ICP) in spite of normal cerebrospinal fluid (CSF) composition, absence of any space occupying lesions, infection and hydrocephalus (Julayanont P et al, 2016). Although IIH has been reported in childhood (Aggarwal A et al, 2017), but epidemiologic studies in this age group are sparse. IIH in pre-pubertal children has distinct characteristics from the adult form (Tepe D et al, 2016). Delayed diagnosis in children usually attributes to the inability in picking subtle visual changes, which can subsequently lead to permanent visual damage. We report two adolescent girls who presented with headache and rapid

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progression of visual impairment. Both the patients were started on medical management along with repeated lumbar punctures initially. However, deteriorating vision was refractory to medical treatment necessitated urgent surgical intervention in the form of lumboperitoneal shunting and resulted in improved visual acuity post operatively in both the patients.

Case Report

Case 1

A 12 years old girl, presented with complaints of headache and blurring of vision for 15 and 7 days respectively. Her headache was diffuse, progressively worsening on straining, coughing, lying down, despite of taking analgesics. Visual deterioration was rather rapid. There were no other associated symptoms and no history of preceding head injury, ear infection, viral illness or use of medications was elicited. Patient's birth history, developmental history and past medical history were otherwise unremarkable.

Physical examination revealed adequate built girl with body weight of 44 kg (between75th to 97th percentile), height of 151 cm (between 50th and 75th percentile) and BMI of 24.1 (between 85th to 97th percentiles). All vital parameters were within normal limits. Systemic and neurological examination was unremarkable except the presence of neck rigidity. On ophthalmologic examination, best corrected visual acuity was finger counting close to face in both eyes**.** Among positive finding, fundus examination revealed bilateral papilloedema. Optical coherence tomography (OCT) demonstrated thickening of nerve fibre layer in both eyes with values of 390 and 376 micrometer in RE and LE respectively. Visual evoked potential showed delay in the P 100 latencies bilaterally, (right eye134 ms, left eye 125ms). Magnetic resonance imaging (MRI) of brain showed distended optic nerve sheaths with tortuosity and indentation of the optic globe without ventricular dilatation (Figure 1). No abnormalities were observed in all major draining sinuses on Magnetic resonance venography (MRV).

Complete blood counts, sepsis screen, iron profile, urine examination, liver and kidney function tests, serum electrolytes, serum cortisol, thyroid functions, antinuclear antibodies, homocysteine, ANA, protein C, protein S and lupus anticoagulant values were within normal limits. Lumbar puncture (LP) revealed clear cerebrospinal fluid (CSF) with opening pressure of 300 cmH_20. CSF biochemistries, staining and culture were negative.

A diagnosis of IIH was made and the patient was started on anti oedema medication in the form of acetazolamide 250 mg thrice a day (Diamox) and low sodium diet. After the diagnostic LP, though symptoms of the patient improved markedly and visual acuity improved to 1/60 in both eyes. But three days afterwards, patient developed severe headache and vision again deteriorated to finger counting close to face. A repeat LP was done and doses of acetalozamide was increased and furosemide (Lasix) was added that lead to improvement in symptoms, with vision improved to 6/36 both eyes. Repeat fundus examination suggested

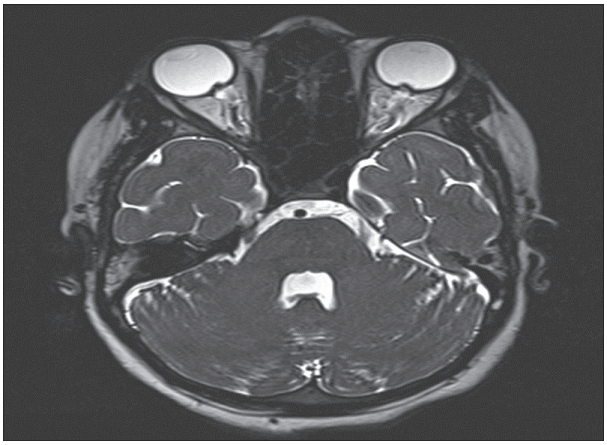

Figure 1: Magnetic resonance imaging (MRI) of brain depicting distended optic nerve sheaths with tortuosity and indentation of the optic globe.

early optic atrophy in both eyes. Single dose of intravenous methyl prednisolone was administered followed by lumboperitoneal shunting. Postoperatively, the patient's vision improved to 6/9. After lumpoperitoneal shunt, a repeat OCT showed significant reduction in retinal nerve fibre thickness owing to resolved papilloedema, thickness reduced to 72 and 80 micrometer in RE and LE respectively. At 6 months, follow up her vision was 6/9 both eyes and remained constant.

Case 2

A 13 years old female patient presented with sudden, painful diminution of vision in both eyes associated with severe, pulsatile headache for twelve days. Diminution of vision was associated with weakness and clumsiness of the right upper arm for five days. There was associated pulsatile tinnitus and fullness of left ear. However, neurological examination was inconclusive. On ocular examination, the visual acuity was 1/60 in both eyes. Among positive findings, there was evidence of mild ptosis and bilateral papilledema (Figure 2). Extra ocular movements were within normal limits. Both Computed tomography (CT scan) and MRI spine revealed normal findings. MRI brain and orbit showed hypoplastic left transverse sinus with indentation by arachnoid granulation at the left transverse sigmoid junction and mild distension of the right optic nerve sheath with indentation on the posterior aspect of the globe. Left optic nerve sheath was not clearly visualized. MR venography has been done to rule out cerebral venous thrombosis and arteriovenous dural fistula, which was found to be negative. CSF opening pressure was >400 cmH_20.

The diagnosis of IIH was made and the patient was started on oral steroids (40 mg per day) and acetazolamide (250 mg TDS). Despite the continued therapy and external lumbar drainage, patient did not show improvement in headache and the visual acuity. After sorting out Neurosurgery consultation emergency lumbar peritoneal shunt was performed. Post-operatively her visual acuity improved drastically to 6/18 and 6/9 in right and left eye respectively, with relief from headache.

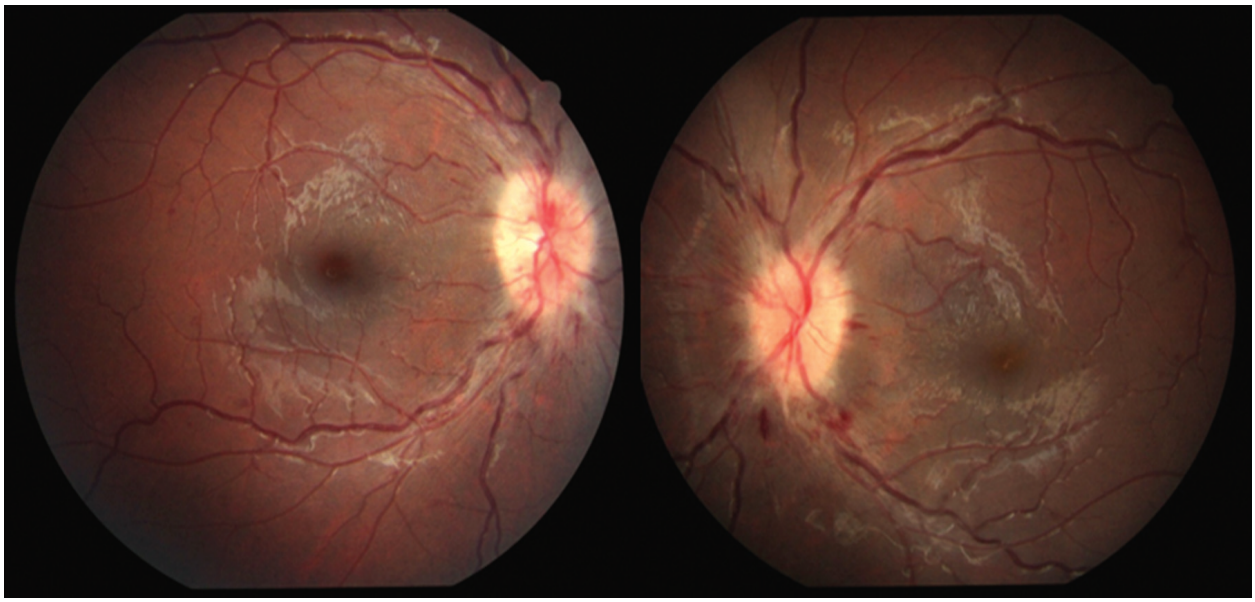

Figure 2: Fundus picture of case 2, depicting bilateral papilloedema.

Discussion

Though the true prevalence of paediatric IIH remains uncertain, its incidence is rising in adolescence due to obesity and improvement in diagnostic modalities (Sugerman HJ et al, 1997). Idiopathic intracranial hypertension may present differently in children than in adults. Recent studies have shown the importance of increased cerebral venous pressures, cerebral venous stenosis and increased venous resistance, collapsed venous structures, increased CSF production and reduced CSF absorption in the pathogenesis of IIH (Biousse V et al, 2012). Thus, MR venography or computed tomography venography is indicated in the workup of such patients to rule out venous thrombosis. Various treatment modalities are documented in literature, such as acetazolamide, furosemide, thiazides, topiramate, corticosteroids (though controversial role), weight reduction, repeated lumbar punctures and CSF diversion surgery (Soler D et al, 1998).

Surgical procedures are reserved to patients who are refractory to medical treatment and in patients with impending visual loss; a CSF diversion procedure can be sight-saving. CSF diversion procedures include lumboperitoneal shunt and ventriculo peritoneal shunts. Non-CSF diversion procedures include optic nerve sheath fenestration, bariatric surgery and venous stenting. Though, optic nerve sheath fenestration is an alternative to shunting, but despite relieving papilloedema on the operated site, it fails to relieve headaches and improvement in visual acuity of fellow eye (Spoor TC et al, 1993). Repeated optic nerve sheath fenestration in failed cases is usually not recommended owing to the less likelihood of improvement and associated risks. Lumboperitoneal shunting is effective in lowering the pressure and decreasing the headache and visual symptoms, in which a catheter is inserted into the subarachnoid space at the level of lumbar spine between two vertebrae and fed around the oblique muscles under the skin into the peritoneum. In present case report, patient was refractory to the medical management, unlike one previous report which showed marked improvement with medical therapy (Aggarwal A et al, 2017). In both the patients, good results were observed after LP shunting. This finding is supported by the previous study done by Burgett et al who described 82% of reduction in headache and 96% resolution of visual symptoms in patients undergoing LP shunt (Burgett RA et al, 1997). Similarly, study observed a 100% resolution in symptoms after LP shunt (Johnston I et al, 1998). However, it is important to realize that earlier the surgery is undertaken, better would be the results. If a delay of 24-48 hrs is anticipated, a lumbar drain should be inserted (Mollan SP et al, 2014).

Among complications of LP shunt; shunt infection, malfunction of the shunt, migration of the shunt and the obstruction of the shunt are the common ones. Over drainage is another concern particularly in those shunts without valves leading to low pressure headache. Hence, prompt diagnosis and aggressive management is the key to better outcome of IIH.

To summarise, although ONSF, VPS, and LPS have documented effectiveness in halting vision loss and are helpful in some visual improvement, all the surgeries are associated with failure rate even after initial success. Hence, enhanced awareness, early diagnosis and prompt treatment are essential to avoid permanent visual damage.

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