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Case of pseudotumor cerebri: A case report

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Abstract
Pseudotumor cerebri or idiopathic intracranial hypertension is a rare condition in children most commonly presenting with headache and vomiting, without any cause identified. Here we report a case of 5year old boy with chronic headache with incidental finding of pseudotumor cerebri without any complications on acetazolamide on follow up. Pseudotumor cerebri is a rare condition but with a morbidity of visual loss and cranial nerve palsy. Hence early diagnosis and management is necessary.

Keywords: Pseudotumor cerebri, partially empty sella turcica

Introduction
Idiopathic intracranial hypertension, also known as pseudotumor cerebri, is a clinical syndrome [1] that mimics brain tumours and is characterised by elevated intracranial pressure (>280 mm Hg in obese or sedated children; 250 mm Hg in nonobese, non-sedated children [2], associated with headaches, papilledema, visual changes, or tinnitus in children with a normal Cerebrospinal fluid cell count and protein count and normal ventricle anatomy and position documented by MRI. There are different hypothesis regarding the etiology of pseudotumor cerebri including increased CSF production or decreased CSF absorption. This condition can potentially cause permanent loss of vision [3]. Hence early diagnosis and treatment is important. Here, we discuss the case of a five-year-old boy who had a persistent headache.

Case Report
A five-year-old male child first born to non-consanguineous marriage presented with complaints of headache for Past 2 years on and off. Headache localized in occipital region on and off more during morning and resolves as the day progress associated with vomiting one episode with no photophobia aggravated while climbing stairs with normal developmental milestones with normal nutrition with no family history of headaches. The child had past history of slip and fall from the first floor three years back for which the child was hospitalized and CT was taken showing minimal depressed fracture in parietal region and was treated conservatively. Physical examination was normal, no cranial nerve palsy, tone normal in all 4 limbs and other systems were normal. Ophthalmologist opinion was obtained, there was no refractive error, B-scan normal no evidence of Papilledema. The blood report showed a normal total Count and hemoglobin and normal peripheral smear study, serum electrolytes and calcium were normal. Neurosurgery opinion was obtained due to the Past trauma history and suggested no intervention. Neurology opinion was obtained and requested for MRI brain. MRI brain showed partial empty sella, bilateral retrobulbar optic nerve Sheaths appear prominent with flattening of post Sclera, bilateral optic nerves showing vertical tortuosity. The child was diagnosed as Pseudotumor cerebri syndrome and was started on tablet Acetazolamide 250 mg 1/2 BD. The patient was asymptomatic and now on follow-up.

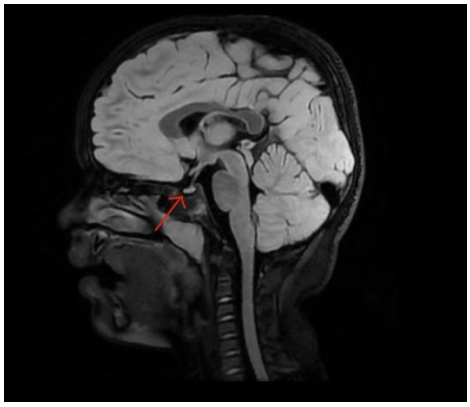

Fig 1: Showing partially empty Sella.

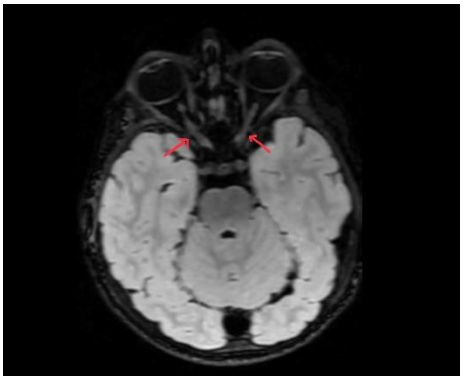

Fig 2: Showing bilateral tortuous optic nerve

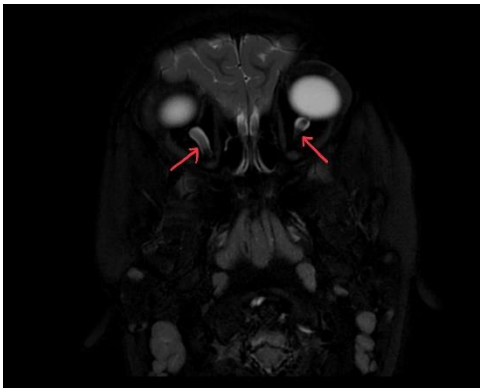

Fig 3: Showing bilateral optic nerve tortousity

Discussion

Pseudotumor cerebri affects both males and females equally in pre-pubertal children with an annual incidence of 0.9 Per 100,000 population [4]. The etiology includes variation in CSF absorption and production, Subtle cerebral edema abnormalities in vasomotor control and cerebral blood flow and venous obstruction, [5] Causes also include metabolic disorders [6, 7], infections drugs and hematological disorders Head injury [8]. The most common presentation in children will be progressive frontal headache (57-87%) that may be associated with visual changes associated with vomiting and visual disturbances. In case of infants, the above condition will characteristically present with bulging anterior fontanel and a "cracked Pot sound"or Mecewen sign (sound produced on percussion near the junction of frontal, temporal, and parietal bones). They may also present with cranial nerve palsy, most common being abducens (CN VI). The evaluation includes history and examination, visual assessment, Neuro imaging including MRI brain with angiogram and venogram [9], lumbar puncture. The MRI will show partially empty Sella turcica [10], flattening of posterior sclera [11], vertical tortuosity of optic nerves [12]. Medical management involves treatment with [13] acetazolamide 10-30 mg/kg/24 hrs [1]. Topiramate is also used rarely. Surgical procedure is done when medical management fails which involves CSF shunting (ventriculo-peritoneal or Lumbo-peritoneal), Optic nerve sheath fenestration, sub temporal decompression. The repeat radiological studies can be performed in refractive cases as it may show evidence of a slow growing tumour or venous obstruction at the time of reinvestigation.

Conclusion

Pseudotumour cerebri is a rare clinical condition yet an important one. The presentation when compared to adults will vary in children. As this condition carries the morbidity of visual loss, cranial nerve palsy, timely diagnosis and management is important as the above morbidity will reduce the quality of life in the patients with untreated Pseudotumor cerebri.

Conflict of Interest

Not available

Financial Support

Not available

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