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Case Report

Transverse Venous Sinus Stenting For Pseudotumor Cerebri Syndrome in a Child Using Recombinant Human Growth Hormone

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Abstract

Objective: To report a rare case of intracranial hypertension with papilledema and visual field loss in a 14-year-old boy receiving recombinant human growth hormone.

Method: Case report.

Results: Lumbar puncture revealed an elevated cerebrospinal fluid opening pressure of 47cmH2O. The hormone was descontinued and acetazolamide 500mg four times daily initiated. However, lumbar pressure was 51cmH2O one week later. Cerebral angiography revealed transverse sinus stenosis. The patient was treated with venous sinus stent implant, with a gradual improvement of the symptoms.

Conclusion: Transverse sinus angioplasty may improve the clinical symptoms and signs of intracranial hypertension in selected patients. Additional research is needed to confirm these findings.

INTRODUCTION

Increased Intracranial Hypertension (IIH), also known as pseudotumor cerebri (PTC), syndrome, is an uncommon disorder of unknown cause, characterized by normal cerebrospinal fluid (CSF) composition and elevated opening intracranial pressure (ICP) (>25cmH_2O in adults and >28cmH_2O in children) documented by lumbar puncture and absence of other possible causes of intracranial hypertension (space occupying lesions, head trauma, encephalitis and meningitis) by neuroimaging or other diagnostic method [1,2]. It is more common in adults, but can occur at any age in chidhood, and may be primary (idiopathic intracranial hypertension) or arise from an identifiable secondary cause.

Usual presenting symptoms of the disorder are a direct consequence of the pressure elevation and include headache, nausea, vomiting, pulsatile tinnitus, blurred vision, strabismus secondary to 6th and 4th cranial nerve palsy and visual field loss of varying degress resulting from papilledema [3,4]. Prepubertal children with primary PTC have an equal sex distribution and less frequent obesity compared with adult patients. Similar to adult patients, children are at risk for the development of permanent visual loss.

Increased CSF, reduced CSF absorption, increased cerebral venous pressure, venous sinus stenosis, increased brain water content, or a combination of these mechanisms have been implicated as a possible mechanism for PTC. Various medical conditions, including systemic diseases, drugs, and vitamin disturbances have been associated with PTC. Of medications associated with PTC, growth hormones, tetracyclines, and vitamin A analogues have been the most often reported [1-4].

Since 1993, many cases of PTC syndrome were reported in children treated with recombinant human growth hormone (rhGH) [5-7]. Cessation of drug is often sufficient for symptom resolution. For patients requiring medical therapy, acetazolamide is the most commonly used medication. However, some patients are refractory to medical treatment alone and develop progressive vision loss, requering surgical intervention, such as optic nerve sheath fenestration, CSF diversion, or venous sinus stenting to control ICP [1,2,7].

This report describes a rare case of a prepubescent boy treated with rhGH for growth failure who presented visual field loss due to refractory PTC syndrome, successfully managed with a single transverse sinus stent.

CASE PRESENTATION

A 14-year-old boy presented with a ten-day history of blurry vision associated with mild headaches and pulsatile tinnitus. He was neither obese nor suffer from renal insufficiency, and was on the 8th week of rhGH treatment on a dose of 2.5 mg daily. On examination, visual acuity was 20/40 in both eyes, with normal color vision and pupillary light responses. Anterior segment and intraocular pressures were unremarkable bilaterally. Fundoscopy revealed extensive fluid around the optic nerve and blurring of the disc margins bilaterally consistent with papilledema (Figure 1A and 1B). Neurological evaluation did not reveal any significant finding. Octopus visual fields revealed inferior defect in right eye and inferonasal defect in left eye. The initial lumbar puncture demonstrated an opening CSF pressure of 47 cmH_2O, but the cell count ($7/mm^3$), protein concentration (33 mg/dl), and glucose concentration (48 mg/dl), were normal, and culture was negative. Brain magnetic resonance imaging (MRI), and angiotomography revealed flattening of posterior sclera, tortuosity of optic nerves and proeminente perioptic nerve sheath, consistent with increased ICP. The rhGH was discontinued and a treatment with acetazolamide (2g daily) was initiated. One week later, his best corrected visual acuity was 20/70 in both eyes and tinittus persisted. Lumbar puncture was repeated and the opening CSF pressure was found to be 51cmH20. Cerebral angiography showed a severe focal stenosis of the right dominant transverse sinus (Figure 2), with a congenitally hypoplastic left transverse and sigmoid sinus. After venography with iodinated contrast material, under general anestesia, intravenous pressure monitoring demonstrated elevated pressure gradiente across the stenosis (20mmHg). A CarotidWall stent (9x50mm; Boston Scientific,Natick, MA, USA) was placed, and it achieved complete dilatation. Repeat pressures demonstrated immediate normalization of the pressure gradiente above and below the stent. Headache and pulsatile tinnitus resolved, and his lumbar CSF pressure dropped to 09 cmH2O on discharge. The patient was medicated with clopidogrel 75mg per day for six months and aspirin 100mg per day for one year. He improved gradually in symptoms and visual fields. After 3 years follow-up, the patient is asymptomatic, but with a little visual field defect is present.

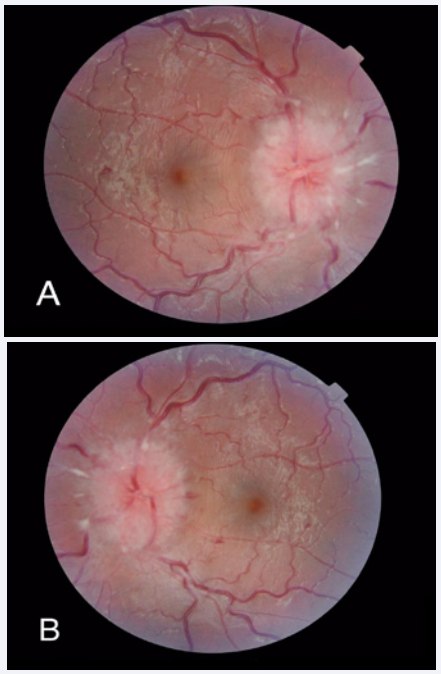

Figure 1 (A): The fundus photographs of the patient at presentation showing bilateral papilledema. (B): The fundus photographs of the patient at presentation showing bilateral papilledema.

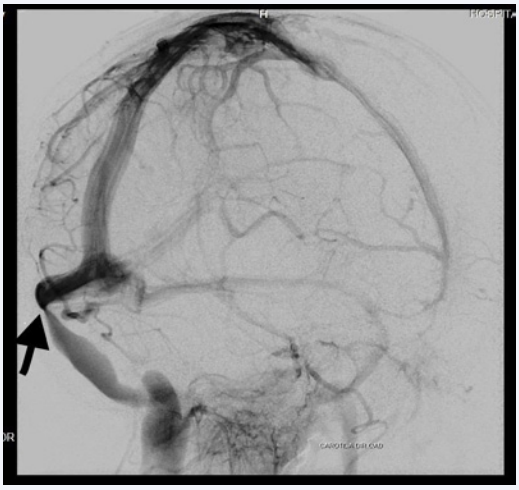

Figure 2 Angiography of the patient revealing severe stenosis of the dominant right transverse sinus (arrow).

DISCUSSION

The diagnosis of PTC is one of exclusion. Ophthalmologic signs of PTC consist of diminished visual acuity, visual field disturbances, pulsatile tinnitus and, papilledema on funduscopic examination. Computerized tomography scanning or MRI of the head should be performed initially to rule out intracranial mass lesions.

The most recently recognized risk factor for PTC is use of rhGH. The incidence of PTC in patients newly started on GH therapy ranges between 0.1% and 0.2% (5). It is dosedependent and occurs mostly within the first months of therapy. As in our case, the majority of reported cases of PTC ocurred at dosages between 0.17 and 0.35mg/kg/day. Symptoms started after one month of therapy.

The natural history of untreated IIH is uncertain. When IIH is due to GH treatment, stopping replacement hormone therapy is often sufficient for symptoms resolution, with a temporary course of acetazolamide, topiramate and steroids used concurrently in most reported cases [5-7]. Patients with IHH who fail, are intolerant to, or are non-compliant with maximum medical therapy and have intractable headache or progressive visual loss may be benefit from surgical intervention. However, these techniques are not free of risk and may lead to complications.

In our patient, the ICP symptoms were refractory and did not respond to acetazolamide.

Although the exact pathophysiology of PTC is unclear, transverse sinus stenoses are observed in high proportion of patients with presumed intracranial hypertension particularly involving one or both transverse sinuses. The belief that the stenoses are the cause, rather than an effect of the increased ICP, has led investigators to recommend stenting of the stenosed sinus as a safe and minimally invasive surgical option for the treatment of the condition. In most studies, stenting of the dural sinus was undertaken when thepressure gradient exceeded or equaled 08 mmHg [8-10]. Transverse sinus stent placement reduced the pressure gradient across the stenosis and improved symptoms in our case.

Ophthalmologic and neurologic evaluation is recommended for children before and during the first few months following the initiation of rhGH therapy. This case support the safety and efficacy of venous sinus stenting in patients with PTC who are refractory or intolerant to medical therapy or with a fulminante presentation. Endovascular treatment can be a good and promising option for treating primary or secondary IHH-associated transverse sinus and sigmoid sinus stenosis, by releasing a stent in the stenosis area.

REFERENCES

1. Ko MW, Liu GT. Pediatric idiopathic intracranial hypertension (pseudotumor cerebri). Horm Res Paediatr. 2010; 74: 381-389.

2. Friedman DI, Liu GT, Digre KB. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. Neurology. 2013; 81: 1159-1165.

3. Park UC, Kim SJ, Hwang JM, Yu, YS. Clinical Features and natural history of acquired thir, fourth, and sixth cranial nerve palsy. Eye. 2007; 22: 691-696.

4. Biousse V, Bruce BB, Newman NJ. Update on the pathophysiology and management of idiopathic intracranial hypertension. J Neurol Neurosurg Psychiatry. 2012; 83: 488-494.

5. Loukianou E, Tasiopoulou A, Demosthenous C, Brouzas D. Pseudotumor Cerebri in a Child with Idiopathic Growth Hormone Insufficiency Two Months after Initiation of Recombinant Human Growth Hormone Treatment. Case Rep Ophthalmol Med. 2016; 2016: 4756894.

6. Reeves GD, Doyle GA. Growth hormone treatment and pseudotumor cerebri: coincidence or close relationship? J Pediatr Endocrinool Metabol. 2002; 15 (suppl 2): 723-730.

7. Rogers AH, Rogers GL, Bremer DL, McGregor ML. Pseudotumor cerebri in children receiving recombinant human growth hormone. Ophthalmol. 1999; 106: 1186-1190.

8. Patsalides A, Oliveira C, Wilcox J, Brown K, Grouver K, Gobin YP, et al. Venous sinus stenting lowers the intracranial pressure in patients with idiopathic intracranial hypertension. J Neurointerv Surg. 2019; 11: 175-178.

9. Miyachi S, Hiramatsu R, Ohnishi H, Takahashi K, Kuroiwa T. Endovascular treatment of idiopathic intracranial hypertension with stenting of the transverse sinus stenosis. Neurointervention. 2018; 123: 138-143.

10. Dinkin MJ, Patsalides A. Venous sinus stenting in idiopathic intracranial hypertension: results of a prospective trial. J Neuro-Ophthalmol. 2016; 0: 1-9.