Optic Nerve Sheath Decompression in Idiopathic Intracranial Hypertension: A Case Report

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Abstract: Idiopathic intracranial hypertension(IHH) is an uncommon disorder characterized by increased intracranial pressure without radiological or laboratory evidence of intracranial pathology. It occurs most commonly in young, obese women, in their third and fourth decades. In this case, visual field test results of a 31-year-old woman with IIH are reported. This patient with amblyopia in her left eye, was treated medically with acetazolamide after being diagnosed with IIH and underwent optic nerve sheath decompression surgery after 8 years.

Keywords: Idiopathic Intracranial Hypertension, Optic Nerve Sheath Decompression.

INTRODUCTION

Idiopathic intracranial hypertension (IIH), previously called as pseudotumor cerebri or benign intracranial hypertension is predominantly seen in young overweight women of childbearing age with an annual incidence of 2.7-19.3/100 000/year(1). Headache is the most common symptom of IIH, and presents in 80%-90% of patients(2). The most popular hypothesis in the pathogenesis of IIH is a decrease in cerebrospinal fluid (CSF) reabsorption. Decreased CSF outflow may be due to arachnoid granulation or possibly dysfunction of absorption in the extracranial lymphatics (3). Typically frontal or bilateral retro-orbital throbbing or pressure headache may be associated with nausea, vomiting, and transient visual obscuration (4).

IIH can cause transient visual obscuration, loss of visual acuity and visual field defects. Transient visual obscuration lasting a few seconds occurs in 68% of patients and is related to papilledema (5). Ischemia of optic nerve head and downward herniation of the parahippocampal gyrus in the tentorial notch are the proposed mechanisms of this symptom. Abducens nerve is the cranial nerve most commonly affected by elevated intracranial pressure. (ICP). Other symptoms are intermittent tinnitus, vertigo, cognitive dysfunction, olfactory nerve involvement, and low frequency hearing loss caused by compression-stretching of cochlear nerve (6-9).

The compression of optic nerve and optic nerve ischemia are the 2 major theories explaining papilledema caused by elevated ICP (10-11). Eventhough papilledema is commonly symmetric, significant asymmetry can be explained by the difference in size of bony optic canals and/or variation of trabecular meshwork in subarachnoid space surrounding the optic disc (12). The swelling of the peripapillary retinal nerve fiber layer, peripapillary hemorrhages, papilla elevation, and congestion of peripapillary vessels are the characteristics of papilledema (13). Visual field (VF) defects are found in 92% of patients at first presentation.After treatment, 50% of patients improve, 28% remain stable and 22% get worse (14). Visual acuity(VA) and color perception are generally preserved in papilledema unless it enters a chronic or atrophic stage. VF defects are related to the optic disc and occur when nerve fiber bundles are damaged at the level of optic disc (3). High CSF pressure, low IOP or systemic blood pressure can cause axoplasmic flow stasis, optic disc edema and resultant intra neuronal ischemia (15). In general, CSF pressure during lumbar puncture in the lateral decubitis position is greater than 25 cm H2O (16). Normal CSF chemistry and cellularity are the mainstays of IIH. Although there are no imaging evidence of ventriculomegaly or a structural cause for increased ICP such as brain parenchymal, ventricular, meningeal or venous sinus abnormality,empty cella turcica, posterior globe flattening, optic nerve head protrusion or stenosis of the transvers sinus, which can lead to venous outflow obstruction can be demonstrated (4-10).

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CASE REPORT

A 31-year-old woman with headache due to raised ICP presented to the hospital on November 2011. She had no history of any other systemic or neural disease. Headache was typically in the form of bilateral frontal or retro-orbital throbbing or pressure sensation, sometimes associated with nausea and transient visual obscuration. There are no photophobia, phonofobia and vomiting. Headache occurred frequently in the mornings. She had bilateral papilledema, diplopia, bilateral intermittent tinnitus, and visual acuity of 20/20 OD and 20/60 OS on Snellen acuity chart. Left eye visual acuity of 20/60 and double vision were due to childhood amblyopia. She was not overweight (weight 70 kg, height 1.65 m, BMI:25,7). Her MRI imaging showed partial empty sella. There was no other pathology in MRI imaging. CSF pressure was 45cm H_2O in the lateral position. CSF biochemistry and cellularity and also basic serum elements were normal. Other reasons causing ICP elevation were negative. She had no history of toxic agent or drug use, obesity, anemia, cerebral venous abnormality, thyroid disease, hypervitaminosis A related to IIH in etiology. CSF chemistry and PCR(polymerase chain reaction) associated with central nervous system infection were negative. Neurological examination was normal except for disc edema and visual changes. The patient was diagnosed with IIH and was treated with 500 mg of acetazolamide orally three times daily for two months. After she received 1000 mg acetazolamide three times daily for two months, the treatment regimen continued three months as 500 mg three times as daily dosage. During the treatment of acetazolamide, headache reduced substantially,visions did not differ, and papilledema improved slightly.

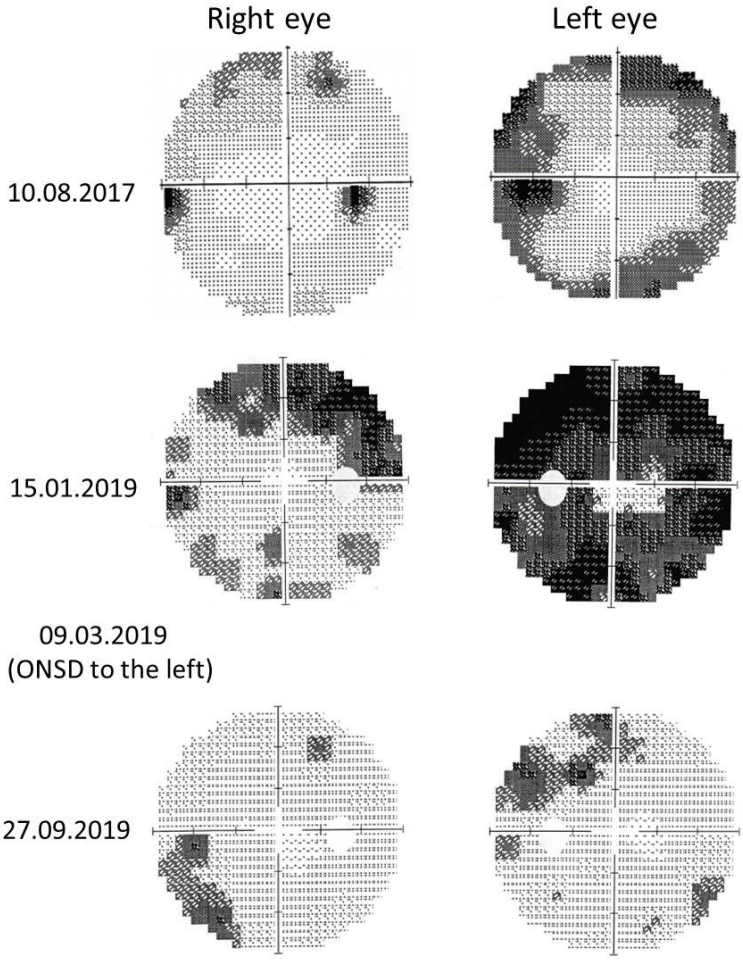

Figure 1: The course of visual fields before and after the surgery (ONSD) in our patient with IIH.

After the medical approach, further intervention was not considered until 2019 because headache was bearable and VFs did not change. On 10 August 2017, automated Carl Zeiss VFs were reliable, MD -3.50dB for OD and MD -10.1 dB for OS and confirmed bilateral defects. Because she became pregnant, VF revealed almost total loss in left eye with high false negative results (MD -6.6 dB OD and -20.1 dB OS with Optovue) and the headache reached intractable level just before the surgery (15 January 2019). The patient underwent ONSD to the left side on 9 March 2019. After performing medial transconjunctival incision, securing and separating the medial rectus muscle, the optic nerve was approached. The optic nerve was dissected from the surrounding tissues and a window was created in the optic nerve sheath. The CSF flowed abundantly into the orbit immediately after incision. There were no intraoperative complications. On the fifth postoperative day, VA was 20/20 in the right eye and 20/60 in the left eye, and complete resolution of papilledema occurred bilaterally. Six months after the operation, headache was at minimal levels and VFs dramatically improved(MD -2.0 dB OD and -2.4 dB OS, with Optovue).

DISCUSSION

As untreated papilledema becomes more chronic, progressive impairment of VA can be expected from a variety of causes, including chronic or atrophic papilledema, chorioretinal folds, macular edema, infarction of the optic disc, subretinal peripapillary hemorrhage and neovascular membrane(10). Risk factors for visual loss are recent weight gain, high-grade papilledema, atrophic papilledema, subretinal hemorrhage, duration of symptoms, pulsatil intracranial noises, degree of headache, pregnancy or opening pressure during lumbar puncture(17). The primary goal of treatment is to prevent visual loss and decrease headache(10). Periodic follow-up by ophthalmologists is essential for reassessing visual status and degree of papilledema(4). Therapy is initiated in the presence of VA or VF loss, moderate to severe papilledema or persistent headache(13). Dietary management and weight loss are strictly recommended(18). Limiting vitamin A consumption and a low tyramine diet may be beneficial(19). A small percentage of patients improve after their diagnostic lumbar puncture. Repeated LPs are sometimes performed in patients with occasional symptom relapses, in pregnant women, or in the setting of rapidly declining vision to temporarily lower the CSF pressure while planning a more aggressive treatment(20). However, the procedure may be painful, technically difficult to perform, and cause a low-pressure headache.

Acetazolamide inhibits carbonic anhydrase enzyme in the choroid plexus, resulting in decreased CSF production and pressure. It is considered for the first-line medication for IIH. The side effects of acetazolamide include paresthesia of extremities, loss of appetite, transient myopia, and metabolic acidosis.

Acetazolamide with a low sodium diet leading to weight loss, a period of 6 months is recommended (21). Topiramate is an anticonvulsant, which has been previously reported to be effective in the treatment of IIH(22). Corticosteroids are useful as an adjunctive treatment in patients with rapid deterioration while arranging surgical procedure(23). When the patient's visual status and optic nerve appearance have stabilized, or when the disease has been in remission for at least six months, ICP-lowering agents may be discontinued. Surgery is considered in cases, including progressive loss of vision despite maximal medical therapy, severe or rapid visual loss at onset,afferent pupillary defect, severe papilledema causing macular edema or exudates(24). Surgical procedures encompass optic nerve sheath decompression (ONSD) and CSF diversion(10). Ventriculoperitoneal and lumboperitoneal shunts are the most widely used methods to divert CSF from the ventricle or subarachnoid space to peritoneal space with the complications of shunt infection, catheter migration, shunt obstruction, and overdrainage (25).

ONSD was first described in 1872 for the treatment of neuroretinitis(26). ONSD has since become a well established surgical procedure for papilledema associated with IIH. The most devastating result of untreated IIH is visual loss. Severe impairment of VA has been reported in up to 10% of patients(14). After medical treatment strategies are exhausted or unable to be tolerated and there is progression of disease, ONSD is a main stay of management to avoid to further deterioration of acuity or VF(27). Surgical complications are diplopia,anisocoria, extraocular movement dysfunction, central or branch retinal artery occlusion, traumatic optic neuropathy, infectious optic neuropathy, transient or protracted blindness(27). Approximately, 50% of patients experience improvement in the nonoperated eye after a single ONSD(28). Filtering effect with local CSF pressure reduction improving the peripapillary circulation,generalised decrease in ICP, scarring of the arachnoid after the procedure protecting the nerve head from the elevated CSF pressure may be the possible mechanisms by which ONSD benefits IIH(28-30). Some studies also reported the improvement of bilateral papilledema and visual function after unilateral ONSD(31). In the early postoperative period, fluid collection adjacent to the dural window site occurs in majority of patients, but this disappears in the late period (32).

The optic nerve compartmentalisation between the intracranial subarachnoid space(SAS) and perioptic SAS has been reported by various authors (33). There may be compartmentalisation at the optic canal in IIH as well. ONSD can lower the pressure within the optic nerve sheath and the intracranial space by allowing CSF flow through the fenestration. CSF flow may not be bidirectional within the optic nerve sheath (33). It is suggested a insufficiency of CSF flow from the intracranial SAS to the SAS of the optic nerve sheath and the anatomical narrowing of the intracanalicular optic nerve sheath SAS could be a contributing factor (33).

We present the VF progress of our patient in this case report (Fig. 1). The patient was initially treated with acetazolamide. Although eight years passed from the first diagnosis of IIH to ONSD, her headache resolved to a great extent, VA and VF unchanged, and papilledema a bit lowered. When she became pregnant VAs did not change, however, her headache reached a intolerable setting, and VFs showed a great depression with unreliable results. After unilateral ONSD was performed, bilateral papilledema completely improved, VF defects disappeared largely, and headache decreased to minimal levels.

As a conclusion, ONSD may be a good alternative treatment in patients who take no benefit in medical treatment.

Conflicts of Interest

There are no conflicts of interest in this case.

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Informed Consent

It was obtained from the patient.

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