Endoscopic Endonasal Optic Nerve Decompression in a Patient With Pseudotumor Cerebri

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Abstract: Pseudotumor cerebri (idiopathic intracranial hypertension) is a syndrome characterized by intracranial pressure elevation and associated signs and symptoms in the absence of a space-occupying intracranial lesion. The most common symptoms are visual loss and headache. Sometimes, surgical therapy is needed in patients who have no apparent response to medical therapy and exhibit a progressive course. Optic nerve decompression is an effective and recommended treatment approach for patients with pseudotumor cerebri in whom visual loss predominates. With the growing experience with endoscopic skull base approaches, this method has begun to be used as an alternative and effective treatment modality. In this study, we aimed to present the outcome of endoscopic endonasal optic nerve decompression and to review the literature on this treatment modality in 2 patients diagnosed with pseudotumor cerebri that was unresponsive to medical therapy and associated with progressive visual loss.

Key Words: Endoscopic endonasal, optic nerve decompression, pseudotumor cerebri

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Pseudotumor cerebri (PTC), also known as benign intracranial hypertension or idiopathic intracranial hypertension, is a clinical entity characterized by intracranial pressure elevation and associated signs and symptoms in the absence of an intracranially located space-occupying lesion. The associated symptoms are due to increased intracranial pressure and papilledema and include headache, nausea, vomiting, and occasionally diplopia.[1–3] The diagnosis is made radiologically by showing the absence of an intracranial lesion and presence of normal-size or small ventricles by computed tomography or magnetic resonance imaging (MRI), an elevated cerebrospinal fluid (CSF) pressure and a normal CSF examination in lumbar puncture (LP), and a normal neurological examination except for the presence of diplopia.[1,2,4–6] This syndrome is usually seen in obese women.[7,8]

Medical therapy is the first choice in PTC. Weight loss, corticosteroids, diuretics, carbonic anhydrase inhibitors (acetazolamide), and serial LPs are the mainstay of therapy.[1,2,9–13] Surgical therapy, on the other hand, is reserved for symptoms that fail to regress or even progress under medical therapy. Surgical approaches include lumboperitoneal shunt, ventriculoperitoneal shunt, foramen magnum–atrial shunt, subtemporal decompression, and optic nerve decompression.[14–23] The latter is a scientifically proven technique that has been applied in the form of lateral orbitotomy, medial orbitotomy, and transconjunctival optic nerve decompression.[20–23] Endoscopic endonasal optic nerve decompression is a novel and effective alternative to surgical therapy, which has largely remained undefined and used in relatively few patients.

In this article, we aimed to assess the effectiveness of endoscopic endonasal optic nerve decompression in 2 patients with PTC with progressive visual symptoms despite optimal medical therapy and to review the relevant literature.

MATERIAL AND METHODS

In this retrospective study, we reviewed the case of 2 patients who were diagnosed PTC with progressive visual deterioration and treated with endoscopic endonasal optic nerve decompression technique. The aim of this study was to discuss and show the effectiveness of this new surgical technique.

Clinical Reports

The first patient is a 31-year-old woman who had occasional headaches for 8 months and had been diagnosed with PTC. The patient had no improvement in her symptoms and even experienced a progressive visual loss despite corticosteroid, diuretic, and carbonic anhydrase inhibitor (acetazolamide) therapy and thus was admitted to our clinic for optic nerve decompression. Neurological and ophthalmologic examination was remarkable for superior hemianopsia at the left eye, bilateral massive papilledema on funduscopy, and a visual acuity ratio of 7/10 in the right eye and 2/10 in the left eye in the visual acuity test (Fig. 1). No lesion was seen on cranial MRI; also, MRI findings were appropriate with small ventricles. An LP was performed, revealing a CSF pressure of 400 mm H_2O. Cerebrospinal fluid biochemical examination did not show abnormalities.

The second patient is a 33-year-old woman who had occasional headaches for 2 years and had been diagnosed with PTC. The patient had no improvement in her symptoms and even experienced a progressive visual loss despite corticosteroid, diuretic, and carbonic anhydrase inhibitor (acetazolamide) therapy and thus was admitted to our clinic for optic nerve decompression. Physical examination revealed bilateral massive papilledema on funduscopy

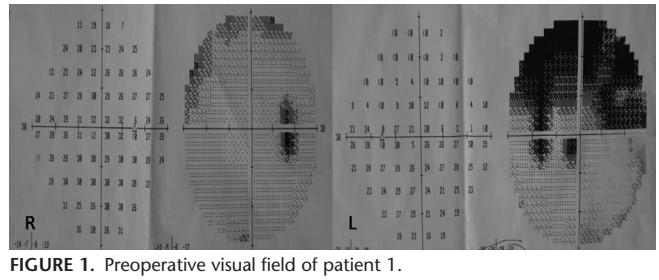

FIGURE 1. Preoperative visual field of patient 1.

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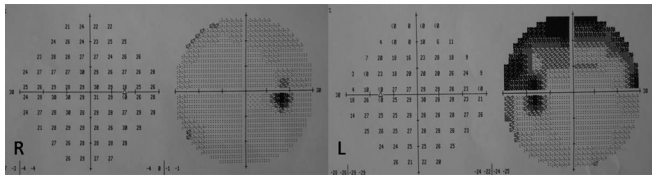

FIGURE 2. Preoperative visual field of patient 2.

and a visual acuity ratio of 9/10 in the right eye and 4/10 in the left eye in the visual acuity test. There were prominent visual field defects in both upper quadrants of the left eye (Fig. 2). No lesion and normal ventricle size were seen on cranial MRI. An LP was performed, revealing a CSF pressure of 340 mm H_2O. Cerebrospinal fluid biochemical examination did not show abnormalities.

Endoscopic endonasal bilateral optic nerve decompression was performed in both patients.

Surgical Technique

Endoscopic endonasal bilateral optic nerve sheath decompression was performed via unilateral right nasal nostril intervention. A 4-mm 0-degree endoscopy device (Hopkins, Karl Storz Endoscope; Karl Storz, Tuttlingen, Germany) was used for the procedure. Middle and superior conchae were pushed laterally, followed by opening of the anterior wall of the sphenoidal sinus and performance of posterior ethmoidectomy. Next, both optic prominences, carotid prominences, and opticocarotid recesses were visualized (Figs. 3 and 4). The thin bony lamella on the intracanalicular part of the optic nerve was opened first with the help of a diamond drill, and decompression was carried out with the help of a microhook and a 1-mm Kerrison rongeur until visualizing orbital fat tissue at the medial and inferior borders of the optic canal on the lateral part. Then, the optic nerve sheath was incised with a microknife crescent to visualize arachnoid herniation and minimal CSF drainage. The same procedure was repeated for the right eye (Figs. 3 and 4). The operation was terminated after filling up the sphenoid bone with absorbable hemostat and pushing the middle concha medially. No C-arm fluoroscopy or nasal tampon was used.

RESULTS

The patients were followed at the ward, and no complication occurred. Their visual examination revealed a visual acuity and field that were unchanged from the preoperative status. Headache and papilledema subsequently improved to some extent, and the patients were discharged on the fourth postoperative day. The headache markedly regressed on the first-month follow-up, but the patients had no prominent change in visual examination. On the third-month follow-up, however, patient 1 reported that she continued to have headache from time to time, whereas patient 2 had no headache. Physical examination at that time revealed markedly regressed papilledema of both patients. The visual acuity level of patient 1 is 7/10 and 4/10 in the right and left eye, and that of patient 2 is 9/10 and 5/10 in the right and left eye, respectively. In addition, the both left eyes manifested a markedly improved visual field (Figs. 5 and 6).

The patients are still in a stable condition at 1-year follow-up.

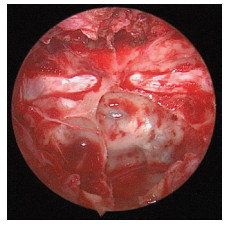

FIGURE 3. Endoscopic view of the optic nerve decompression.

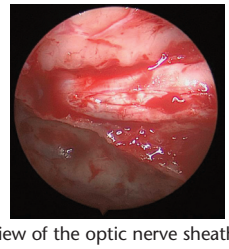

FIGURE 4. Endoscopic view of the optic nerve sheath decompression.

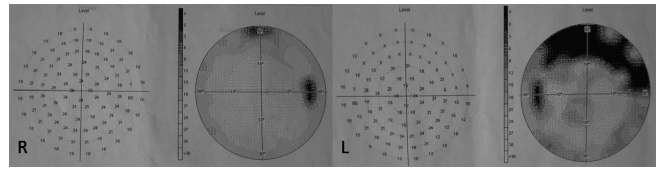

FIGURE 5. Postoperative visual field of patient 1 after 3 months showing significant improvement in the left eye.

DISCUSSION

Pseudotumor cerebri is a clinical entity of unclear etiology and pathophysiology that is characterized by increased intracranial pressure and associated symptoms due to a defect in CSF absorption or circulation.[24,25] Not all patients with PTC need surgery. The generally accepted approach in treatment of the disorder is recommending medical therapy for a maximum period as tolerated before a surgical invasive procedure is attempted. Many patients with this condition can be followed up for many years by putting into practice weight loss, medical therapy, and serial LPs.[3,4] However, surgery may be eventually necessary in patients with continued symptoms or progressive visual loss. Some studies have reported that 12% to 25% of patients followed by medical therapy require surgical operation.[26,27]

Currently, many surgical methods are used in the surgical management of PTC. Although lumboperitoneal shunt applications and effective techniques are commonly used, they are not preferred as first-line therapy owing to subsequent need of multiple revision surgeries and higher complication rates.[15,28,29]

Intraorbital optic nerve sheath decompression is an option with proven effect that has been long used. Many techniques have been defined so far, and transcranial or transorbital approaches have been preferred. The generally accepted and used approach is the medial transconjunctival approach. A review of 7 retrospective studies using this technique has revealed that 423 eyes (on 252 patients) underwent optic nerve sheath decompression with resulting 50% improvement in visual acuity and 72% improvement in visual field, whereas 11% of patients experienced a deterioration of visual function, and 12% required reoperation.[21,23,30–34] The complications associated with this approach had a rate of 4.8% to 45% and included orbital hemorrhage, retinal artery occlusion, sudden intraocular pressure increase, iris sphincter paresis, transient diplopia, and accommodation failure.[20,35,36]

Endoscopic endonasal optic nerve decompression is a novel and minimally invasive treatment option. It has been reported to be successful in cases with traumatic optic neuropathy.[37] This

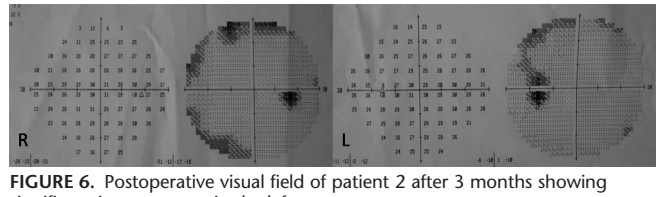

FIGURE 6. Postoperative visual field of patient 2 after 3 months showing significant improvement in the left eye.

approach possesses a number of advantages over other techniques including a better surgical field view, lower morbidity, preservation of olfactory function, short operative time, no externally apparent surgical scar, and better surgical results.[38–40]

In PTC patients, however, endoscopic endonasal optic nerve decompression has been only rarely tried. Compared with transorbital approaches, this approach provides a wider working field, an easier access to the entire intracanalicular optic nerve from optic chiasma to orbital apex and more chance for intervening bilateral optic nerves simultaneously, and low complication rates.[4] There are only a few patients treated with this approach in the literature. Patrocinio et al[1,41] and Gupta et al[1,41] reported that they achieved fair results with unilateral optic nerve decompression. Koc et al[4] reported good outcomes with bilateral optic nerve decompression in 2 patients. We also preferred bilateral optic nerve decompression in 2 patients with loss of visual acuity and visual field in the left eye. We are of the opinion that decompression of the contralateral optic nerve without the need of an extra incision or additional morbidity would ease CSF circulation and exert a prophylactic effect on the eye without symptoms.

CONCLUSIONS

In patients with PTC, endoscopic endonasal optic nerve decompression offers a novel, effective, and minimally invasive treatment option. It should be remembered that this approach must be performed by surgeons with adequate experience in endoscopic skull base operations to avert optic nerve and ophthalmic artery injuries. Literature data on this approach are limited to a few case reports, making a comparison with other modalities impossible. With the fair results it has achieved, however, it still shows promise for treatment of patients with PTC.

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