Case Report

Endoscopic Optic Nerve Sheath Fenestration for Treatment of Papilledema Secondary to Intracranial Venous Hypertension: Report of Two Cases

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Objective: *To determine the safety and efficacy of endoscopic optic nerve sheath fenestration (ONSF) for the reversal of papilledema in intracranial venous hypertension.*
Material and Method: *A retrospective chart review was performed on two consecutive patients who underwent endoscopic ONSF. Presenting symptoms, neuro-ophthalmological work-ups, including visual acuity (VA), visual field charting (VF), optical coherence tomography (OCT) and MRI as well as magnetic resonance venography (MRV) were recorded. Cerebrospinal fluid pressure was also measured preoperatively in both individuals. Visual improvement was assessed by comparing with preoperative ophthalmological findings.*
Results: *This report is the first endoscopic ONSF study focusing on treatment of papilledema resulting from intracranial venous hypertension (tumor compressing transverse sigmoid junction in the first patient and venous sinus stenosis in the second patient). ONSF was performed on both sides of the first patient and on the right optic nerve of the second patient with showing reduction of papilledema on both eyes. Papilledema was improved in both individuals. Vision improved more in the first patient than in the second whom had pre-existing optic nerve atrophy.*
Conclusion: *Endoscopic optic nerve sheath fenestration is an effective and safe procedure to revert visual loss or to stabilize vision in patients presenting with visual loss caused by intracranial venous hypertension.*

Keywords: *Cerebral venous hypertension, Intracranial hypertension, Optic nerve sheath fenestration, Optic decompression, Endoscope*

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Secondary intracranial hypertension is the terminology used when an underlying cause of increased intracranial pressure is detected such as cerebral venous thrombosis or a space-occupying lesion compressing venous outflow. Forty percent of patients with cerebral venous thrombosis may have the manifestation mimicking idiopathic intracranial hypertension[1,2]. Idiopathic intracranial hypertension (IIH) is a condition of increased intracranial pressure of unknown etiology. The most common symptoms are headache, visual loss, transient visual obscuration and diplopia[3]. Medical strategies include weight reduction, serial lumbar puncture, and diuretic or oral acetazolamide to reduce intracranial pressure (ICP). Surgical treatment should be considered when visual loss or headaches progress despite maximum medical therapy.

Optic nerve sheath fenestration (ONSF) is an effective method in patients with IIH who have progressive visual loss from chronic papilledema[4-7]. It results in decreased optic disc swelling as well as an improvement in visual acuity and visual field abnormalities. This treatment was initiated in 1872 by DeWecker[8]. Galbraith and Sullivan first described the medial transconjunctival microsurgical approach to ONSF in 1973[9]. Tse et al then have popularized the lateral orbitotomy approach[10].

Since experiences with endoscopic skull base approaches have been gained, transnasal endoscopic

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optic nerve decompression either with or without nerve sheath opening has begun to be used as an alternative treatment modality in IIH[11-13]. This present study aimed to present the outcomes of endoscopic endonasal optic nerve sheath fenestration in two patients diagnosed with secondary intracranial hypertension resulting from cerebral venous hypertension.

Material and Method

The study was approved by the institutional review board of Prasat Neurological Institute (PNI). The medical records of both patients who had endoscopic optic nerve sheath fenestration at PNI were retrospectively reviewed.

Case 1

A 50-year-old Thai female presented with gradually decrease in visual acuity for six months. The patient had a pre-operative visual acuity (VA) of 20/30 in both eyes. Fundoscopy revealed bilateral papilledema (Fig. 1). There was no other abnormal neurological deficit. MRI scan was obtained and demonstrated multiple small meningiomas of left clinoid, left parasagittal region, left frontal and parietal convexities and small meningioma at right transverse sigmoid junction narrowing the right transverse sinus (Fig. 2). Size of each tumor was less than 1 cm OCT confirmed the clinical features of optic nerve papilledema (Fig. 3). Lumbar puncture measured an elevated opening pressure (30-cm water) with normal CSF studies. Medical treatments using carbonic anhydrase inhibitor and diuretics were given for three months, but without improvement of VA. Tumor at the sigmoid sinus was not considered to be removed due to the high chance of obliterating the sinus instead of reducing the venous hypertension. Therefore, endoscopic optic nerve fenestration using one nostril technique was proposed to the patient.

Case 2

A 29-year-old African female presented with chronic headache and decrease in VA for two years. The patient had a pre-operative visual acuity (VA) of 20/40 in both eyes. Fundoscopy revealed bilateral papilledema and optic nerve atrophy (Fig. 4). OCT was obtained and demonstrated decreased nerve fiber layer (Fig. 5). MRI was unremarkable. However, Magnetic Resonance Venography (MRV) showed stenosis of bilateral transverse sinuses (Fig. 6). Lumbar puncture measured an elevated opening pressure (33-cm water) with normal CSF studies. After discussing with the patient about the treatment modalities either venous sinus stent or ONSF, the patient underwent the ONSF approach.

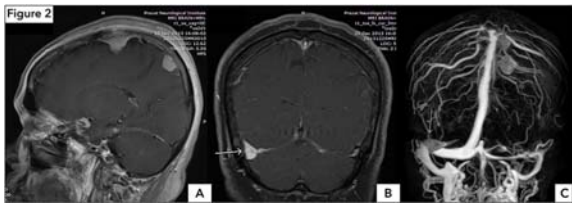


Fig. 2 Sagittal and coronal MRI with gadolinium enhancement demonstrate multiple small meningioma (A&B). Tumor at right transverse sigmoid junction compresses the venous sinus and creates the high intracranial pressure (C).

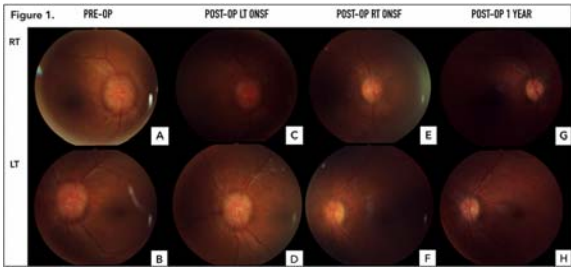


Fig. 1 (A&B) demonstrate the bilateral papilledema of the first patient. (C&D) show the fundoscopy of right and left eyes, one-month after left endoscopic ONSF. However, right optic nerve still revealed papilledema. Therefore, the patient underwent right endoscopic ONSF. (E&F) reveal decreased degree of papilledema at one-month after right endoscopic ONSF. (G&H) show normal bilateral optic fundi.

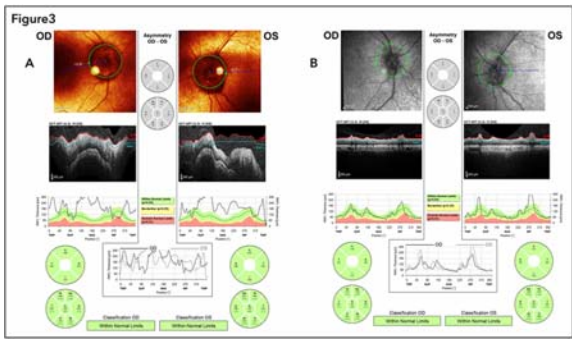


Fig. 3 The preoperative optic coherence tomography (OCT) confirms the clinical features of bilateral papilledema (A). The postoperative OCT (B) shows that the thickness of both optic nerve decreased to normal.

Surgical technique

Endonasal endoscopic ONSF was performed using one nostril technique in the first patient (case 1), on the left optic nerve first. After observing for one month and found that there was no improvement of right optic nerve papilledema, right optic nerve was operated (Fig. 1). In the second patient (case 2), endoscopic ONSF was performed on the left optic nerve using bi-nostril technique due to narrow space. A 4-mm 0-degree endoscopy device (Hopkins, Karl Storz Endoscope, Germany) was used for the procedure. Middle and superior turbinate were pushed laterally, followed by uninectomy, anterior and posterior ethmoidectomy. Sphenoidotomy was performed for orientation. The thin lamina papyracea was removed and optic canal was drilled. Optic nerve sheath was incised with a microknife to visualize arachnoid herniation with minimal CSF drainage (Fig. 7).

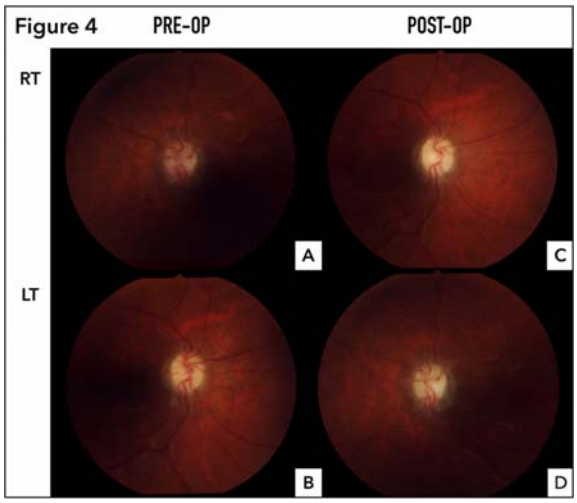


Fig. 4 Fundoscopy of left and right eyes reveal bilateral papilledema (A&B). One month after the operation, both papilledema improved (C&D).

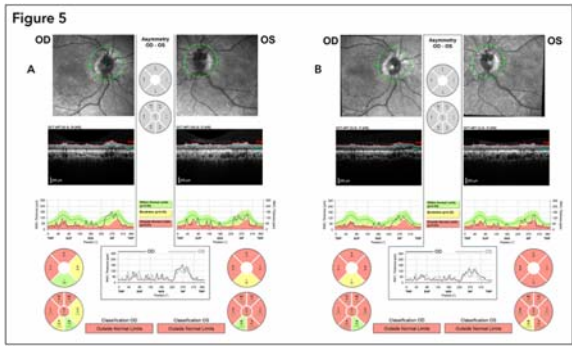


Fig. 5 The preoperative OCT (A) shows the decreased nerve fiber layer which is similar to the postoperative OCT (B). Both OCTs confirm the evidence of reduced papilledema.

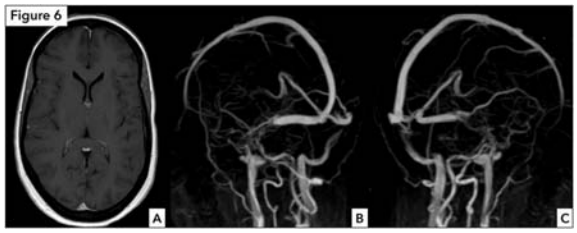
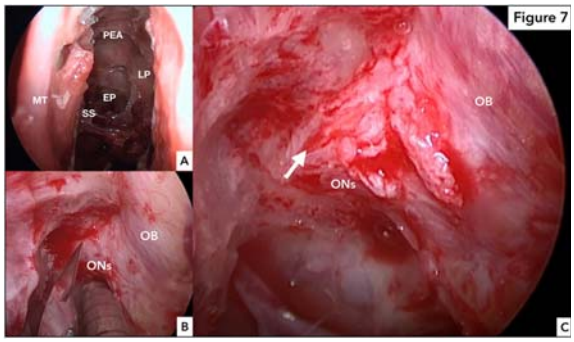


Fig. 6 In the second patient, MRI scan reveals no ventriculomegaly (A). MRV demonstrates the bilateral venous sinus stenosis (B&C).

Results

Headache and diminished vision were the two presenting complaints in both patients diagnosed with cerebral venous hypertension. The two also showed marked relief from the headache and papilledema on the first-month follow-up. They had shown improvement in visual acuity since the third postoperative day. The visual acuity of the first patient returned to normal, whereas the second patient had postoperative VA of 20/30, 20/30. Vision improved more in the first patient than in the second who had pre-



EP = Posterior ethmoidal sinus; LP = Lamina papyracea; MT = Middle turbinate; OB = Medial wall of orbit; ONs = Optic nerve sheath; PEA = Posterior ethmoidal artery; SS = Sphenoid sinus

Fig. 7 Middle and superior turbinate were pushed laterally, followed by uninectomy, and anterior and posterior ethmoidectomy. Sphenoidotomy was performed for orientation (A). The thin lamina papyracea was removed and optic canal was drilled. Optic nerve sheath was incised with a microknife (B) to visualize arachnoid herniation and minimal CSF drainage (C).

existing optic nerve atrophy. Postoperative OCT at one-month demonstrated the decreased nerve fiber layer and reduced degree of papilledema (Fig. 3 and 5). There were no postoperative complications.

Discussion

It has been shown that the lack of even one functioning transverse sinus may lead to venous hypertension and intracranial hypertension[14-16]. We still know little about pathogenesis of venous hypertension creating the intracranial hypertension. Some may propose that the intracranial hypertension is the result of a decreased CSF absorption (without ventriculomegaly) secondary to venous hypertension. Visual loss in cerebral venous thrombosis is multifactorial and includes pressure effects of transmitted ICP on the optic nerve and vascular compromise to the optic nerve and the rest of the optic pathway due to thrombosis[1]. Selection of treatment modalities for venous sinus stenosis resulting in papilledema is still debated. Endovascular venous sinus stenting was recently introduced and could be an optional treatment. VP or LP Shunt procedures may be other alternative surgical intervention[17], which can be associated with shunt revision due to shunt failure. The two main proposed mechanisms of ONSF in improving visual function include obliteration of the subarachnoid space surrounding the optic nerve by fibroblast proliferation, preventing CSF pressure transmission distal to the operative site[18] and another potential explanation is the creation of a dural fistula which allows egress of CSF from the operative site[19]. Although the pathophysiologic mechanism by which ONSF improves visual function is not settled, there is evidence on the efficacy of ONSF in improving or preserving vision in patients with intracranial hypertension due to other causes like IIH or cerebral venous thrombosis[1,2,16,20-22]. Therefore, ONSF was used in the treatment of IIH or intracranial hypertension by different approaches, usually with orbitotomy. The significant complications include accommodation paresis, iris sphincter paresis, and central retinal artery occlusion[23-25]. So as to overcome such complications, we would recommend the technique of endoscopic ONSF for treating intracranial hypertension due to cerebral venous hypertension. To our knowledge, this paper may be the first report mentioning about the treatment of papilledema in intracranial venous hypertension due to venous sinus stenosis using the endoscopic ONSF.

It is also interesting that the second patient experienced the reduction of papilledema in both operated and non-operated eyes. Even though the mechanism for this contralateral nonsurgical effect is not well understood, it could be related to the reduced intraoptic nerve pressure[26] which allows axoplasmic flow to begin again. In conclusion, endoscopic ONSF is an effective treatment for papilledema in patients diagnosed with intracranial venous hypertension.

What is already known on this topic?

Optic nerve sheath fenestration is an effective method in patients with IIH who have progressive visual loss from chronic papilledema. This treatment was described using the medial transconjunctival microsurgical approach or the lateral orbitotomy approach. Transnasal endoscopic optic nerve decompression either with or without nerve sheath may be used as an alternative treatment modality in idiopathic intracranial hypertension.

What this study adds?

The authors presented the outcomes of endoscopic endonasal optic nerve sheath fenestration in two patients diagnosed with secondary intracranial hypertension resulting from cerebral venous hypertension. To our knowledge, this paper may be the first report mentioning about the treatment of papilledema in intracranial venous hypertension due to venous sinus stenosis using the endoscopic ONSF.

Potential conflicts of interest

None.

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การผ่าตัดเปิดปลอกหุ้มเส้นประสาทตาด้วยกล้องเอ็นโดสโคปในรายที่มีปัญหาจอประสาทตาบวมจากการที่มีภาวะความดันสมองสูงที่มีสาเหตุจากหลอดเลือดดำใหญ่ในสมองตีบ

วุฒิพงษ์ ฐิรโฆไท, พัชรพิมพ์ มัศยาอานนท์, จิรพร สุวรรณสัญญา, เอื้ออังกูรย์ สิทธิมงคล, บุญสาม รุ่งภูวภัทร

วัตถุประสงค์: *เพื่อประเมินประสิทธิภาพการผ่าตัดภาวะจอประสาทตาบวมด้วยกล้องเอ็นโดสโคปที่มีภาวะความดันสมองสูงอันเกิดจากเส้นเลือดดำใหญ่ในสมองตีบ*

วัสดุและวิธีการ: *ผู้ป่วยจำนวน 2 ราย ที่มีภาวะจอประสาทตาบวมจากภาวะความดันสมองสูง สาเหตุจากการที่มีภาวะความดันในหลอดเลือดดำใหญ่สมองสูง ได้รับการตรวจสายตา (VA), ลานสายตา (VF), optical coherence tomography (OCT), MRI และ MRV นอกจากนี้ยังมีการวัดความดันสมองก่อนการผ่าตัดทั้งสองรายด้วยการเจาะน้ำไขสันหลัง ผลที่ได้จากการรักษาใช้การประเมินผลทางจักษุวิทยาด้วยการตรวจวัดสายตา ลานสายตา และ OCT*

ผลการศึกษา: *เป็นการศึกษาครั้งแรกในการรักษาจอประสาทตาบวมที่มีภาวะความดันสมองสูงจากเนื้องอกสมองกดทับบริเวณ transverse sigmoid junction ในผู้ป่วยรายแรกและภาวะ venous sinus stenosis ในผู้ป่วยรายที่สองโดยทำการผ่าตัดเปิดปลอกหุ้มเส้นประสาทตาทั้งสองข้างในผู้ป่วยรายแรก ส่วนผู้ป่วยรายที่สอง ผ่าตัดเฉพาะเส้นประสาทตาด้านขวา จากการผ่าตัดผู้ป่วยทั้ง 2 ราย มีการลดลงของภาวะจอประสาทตาบวม พบว่าผู้ป่วยรายแรกสามารถมองเห็นภาพได้ชัดเจนมากกว่า เนื่องจากผู้ป่วยรายที่สองมีปัญหาจอประสาทตาเสื่อมก่อนผ่าตัด*

สรุป: *การผ่าตัดเปิดปลอกหุ้มเส้นประสาทตาด้วยกล้องเอ็นโดสโคปเป็นวิธีที่มีประสิทธิผลและมีความปลอดภัย เพื่อรักษาการมองเห็นให้ดีขึ้นอันมีสาเหตุจากภาวะความดันสมองสูง*