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CLINICAL REPORT

# Idiopathic Intracranial Hypertension and Endoscopic Optic Nerve Sheath Fenestration

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**Abstract** Idiopathic intracranial hypertension (IIH), also called as benign intracranial hypertension is a disorder, which is considered benign in its course except its' ill effects on vision. Ocular findings in IIH such as papilledema, macular changes, retinal micro haemorrhages, cotton wool spots and tortuosity of vessels are the prominent features in funduscopy examination in these patients. Papilledema is a hallmark feature for evaluation of response to treatment. Ophthalmological rescue is a primary goal of management of idiopathic intracranial hypertension. Among the treatment options described in literature, optic nerve sheath fenestration is a minimally invasive endoscopic technique for the rescue of vision. We present this case-report, which will help ophthalmologists and the surgeons to determine the significance of the funduscopy changes after optic nerve sheath fenestration and help in decision making.

**Keywords** Idiopathic intracranial hypertension · IIH · Optic nerve fenestration · Optic nerve sheath decompression procedure · ONSD · Visual deterioration with papilledema

## Introduction

Idiopathic intracranial hypertension (IIH) is a disorder resulting from raised cerebrospinal fluid pressure in the absence of an intracranial mass lesion or ventricular dilatation with normal spinal fluid composition, in an alert and awake patient. Visual function tests are an integral part of neurological examination for diagnosing and monitoring IIH patients and may reveal papilledema, macular involvement, sub-retinal haemorrhage, visual field and nerve fibre bundle defects [1]. We report a case highlighting the potential resolution of fundus changes following endoscopic optic nerve sheath fenestration and discussing the significance of these changes.

## Case-Report

A 23-yr-old healthy girl having insignificant past and family history presented to our OPD complaining of throbbing headache and insidious bilateral deterioration of vision for 6 months without any history of pain or photophobia. Examination revealed full range of motion extra ocular movements (EOM) with bilateral diminished visual acuities (VA), which were perception of face-close hand-movements on right and 6/24 in the left, relative afferent papillary defect (RAPD) with swinging light reflex test, optic atrophy in funduscopy in right and chronic papilledema in left with bilateral prolonged p100 latency in visual evoked potentials(VEP) suggesting of anterior visual pathway defect. Magnetic resonance imaging(MRI) of the brain with venography showed hypoplasia of right and short segmental narrowing of left transverse sinuses without hydrocephalic features, flattened posterior globes, and tortuous optic nerves in both eyes. CSF opening pressure (Lumbar puncture) was 50 cm of water, with normal cellular and biochemical constituents in analysis. Diagnosis of IIH was made considering modified Dandy's criteria, patient was admitted, prescribed with 500 mg of acetazolamide BD. Endoscopic-optic-nerve-sheath-fenestration

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(ONSF) was planned on the right side for visual rescue and carried out. Two fenestrations each of the size of 1 × 2 mm were created over optic nerve sheath, one just behind annulus of zinn and second near the base of skull. Acetazolamide administration continued with 5-day course of intravenous pulse dose methylprednisolone therapy as given in optic neuropathy.

After one week, headache relieved without visual improvement signs though funduscopy exhibited resolution of haemorrhages. Over next few weeks she persistently improved and became neurologically asymptomatic at three months post-op with stabilization of VA without improvement/worsening in the right and improvement of VA to 6/9 in left and 3-month post-op funduscopy revealed right sided optic atrophy with improvement on the left. Serial examinations and fundus photography over next of 3 months showed no deterioration. Fundus photography images at presentation depicted severe papilledema with nerve fibre layer haemorrhages and vessels tortuosity in left eye (Fig. 1a) and right eye (Fig. 1b). Images at 1 week post-treatment showed mild papilledema with resolution of haemorrhages in left eye (Fig. 2a) and right eye (Fig. 2b) and images at 3 months post-treatment showed resolution of papilledema in left eye (Fig. 3a) and secondary optic atrophy in right eye (Fig. 3b). (Table 1).

## Discussion

IIH is multifactorial and characterized by severe headache, nausea, vomiting, transient visual obscuration and diplopia, and no underlying cause explaining the symptoms/signs attributable to increased intracranial-pressure(ICP) evidenced during lumbar puncture with normal cerebrospinal fluid (CSF) composition; no imaging evidence of ventriculomegaly or a structural cause for increased ICP such as a brain parenchymal, ventricular, meningeal or venous sinus abnormality; and no other identified cause of intracranial hypertension, such as use of certain medications [2].

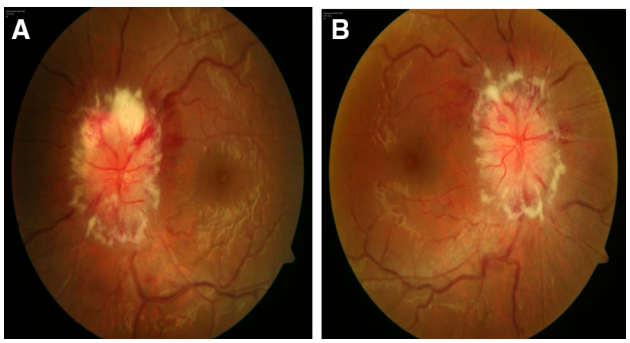


**Fig. 1** Fundus photography images at presentation. Left eye (**a**) and Right eye (**b**)

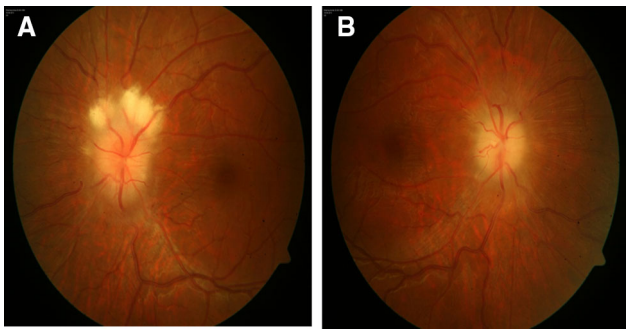


**Fig. 2** Fundus images at 1-week post-treatment. Left eye (**a**) and Right eye (**b**)

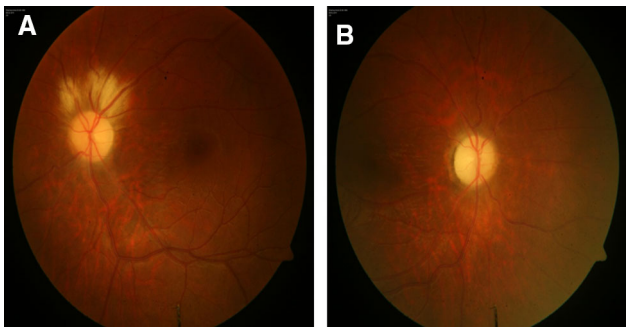


**Fig. 3** Fundus images at 3 months post-treatment. Left eye (**a**) and Right eye (**b**)

Papilledema, optic disc oedema, due to increased intracranial pressure is the hallmark sign of IIH. Conventionally, the term papilledema is reserved for the cases of optic disc oedema when the swelling is due to raised ICP and does not arise from local optic nerve processes such as inflammation, extrinsic compression or ischemia. Raised CSF pressure is transmitted through the optic canal into the intra-orbital optic nerve sheath to exert effects on the axons of the optic nerve at the point of exit from the eye through the lamina cribrosa. Fast and slow axoplasmic transport is interrupted by the raised pressure, resulting in nerve fibre swelling at the nerve head. At the optic nerve head and peri-papillary region, some features of central retinal vein obstruction are typically seen. These include dilatation of the pre-papillary retinal capillaries and nerve head hyperaemia, dilation of retinal veins, peri-papillary haemorrhages and cotton wool spots. These are the consequences of compression of the central retinal vein, initially by the dilated axons within the pre-laminal optic nerve head and later by pressure from the CSF on the central retinal vein as it crosses the subarachnoid space to exit the optic nerve 1 cm behind the globe [3–5]. The changes of papilledema may sometimes be highly asymmetrical or even monocular and chronic raised intracranial pressure needs to be

**Table 1** Showing the course of disease

| Time of assessment | Non-visual symptoms | Fundus examination | | Visual acuity | |
|---|---|---|---|---|---|
| | | RE | LE | RE | LE |
| At presentation | Headache | Severe Papilledema, haemorrhages, tortuous vessels | Severe Papilledema, haemorrhages, tortuous vessels | HM | 6/24 |
| 1 week post-treatment (Rx) | Nil | Mild papilledema, resolution of haemorrhages | Mild papilledema, Resolution of haemorrhages | HM | 6/24 |
| 3 months post-Rx | Nil | Secondary optic atrophy | Resolution of papilledema | HM | 6/9 |

included in the differential diagnosis of unilateral cases [6–8]. Structural factors of the optic disc may play a role in the development of or lack of papilledema in patients with IIH resulting in asymmetrical papilledema [9]. Routine clinical observations, serial visual field charting, direct imaging of the optic nerve head, serial fundus photography, ultrasound B-scanning and optical coherence tomography; can all be used in monitoring IIH patients. Colour fundus photography has proven to be an excellent way to record the fundus findings [10].

Serial disc photography is also valuable for long-term monitoring. Peri-papillary retinal nerve fibre layer imaging with ocular computed tomography is also being increasingly used [11].

Primary indications for the treatment of IIH are intractable headache and vision loss. Surgery is considered when there is progressive loss of vision despite maximal medical therapy; severe papilledema causing macular oedema, exudates and severe or rapid visual loss at onset [12, 13]. Surgical procedures used for the treatment of visual loss include ONSD and CSF diversion procedures. Outcomes from optic nerve sheath decompression (ONSD) are better documented and appear to be superior to other surgical techniques for managing visual loss [14] whereas diversion and perhaps stent placement procedures proves helpful in managing headache.

In this case we had done optic nerve sheath fenestration (ONSD) as a rescue procedure. An interesting finding to be observed in this case is that though we had taken initial steps of performing ONSD in the worse eye at first in order to take a guarded approach, patient showed improvement in both eyes. This indicates the effective role of the ONSD procedure in one eye to effect and act as a rescue for the other eye by reducing the pressure. These effects of unilateral ONSD have also been documented in literature. In this case, we emphasize the role of optic nerve sheath fenestration (ONSD) procedure, which creates 2 small windows in the optic nerve sheath compared to the traditional technique of longitudinal incisions in the optic nerve sheath.

In our case, serial disc photography revealed resolution of haemorrhages after 1 week and disappearance of exudates after 3 months of optic nerve sheath fenestration, which was notable, with no improvement or deterioration of vision in the right eye as optic atrophy had already set in. However, the improvement in VA seen in the left eye indicates the efficacy of optic nerve fenestration as an ONSD procedure with surgical creation of 2 windows in one of optic nerve sheaths. The improvement of symptoms and visual stabilization was clinically sufficient to avert the need to proceed with a contralateral ONSD.

## Conclusion

The mechanism for this contralateral surgical effect is not certain and could be related to decreased intra-sheath CSF in both optic nerves after unilateral ONSD, regression toward the mean, spontaneous improvement, or simply better patient compliance with maximum medical therapy after unilateral surgery. However, it can be very well emphasized that optic nerve fenestration can act as an effective rescue procedure in idiopathic intracranial hypertension.

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**Compliance with Ethical Standards:**

**Conflict of interest** There exist no conflicts of interests among the authors, and there are no financial disclosures to be made.

**Ethical approval** It is a case report and is being reported after the management of the case. Hence, it is exempted from institutes' ethical requirements.

**Informed consent** Informed detailed consent has been taken from the patient for the surgery and management at every step. Informed consent has also been obtained from the patient for using clinical data for publication and research purposes on the conditions of anonymity.

Hence, it was ensured that patients name is not disclosed in the text or figures.

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