



## UNILATERAL PAPILEDEMA IN ASYMPTOMATIC ELDERLY MALE: AN UNCOMMON PRESENTATION OF IDIOPATHIC INTRACRANIAL HYPERTENSION

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### ABSTRACT

Asymptomatic unilateral papilledema is an atypical presenting sign of Idiopathic intracranial hypertension (IIH). We report a rare case of a 65 year old elderly male of medium built who was asymptomatic and reported for the routine eye check up. Ocular examination revealed optic disc swelling in the left eye. Subsequent contrast enhanced Magnetic Resonance Imaging of the brain and orbit revealed features of widened and partially empty sella. Lumbar puncture revealed an elevated opening cerebrospinal fluid pressure of 268 mm of water thus diagnosis of IIH was made. He was subsequently treated with oral tablet acetazolamide. Therefore in cases of unilateral disc edema where patient is asymptomatic, diagnosis of IIH should always be kept in mind irrespective of the age group and gender of the patient.

**KEYWORDS :** Idiopathic intracranial hypertension; Papilledema; Pseudotumor cerebri

### INTRODUCTION

IIH is a disorder of unknown etiology that mainly affects women who are fat and are of childbearing age<sup>1</sup>. Idiopathic Intracranial Hypertension (IIH) is also termed as benign intracranial hypertension so as to differentiate it from secondary intracranial hypertension which is as a result of neoplastic malignancy. However IIH is not a benign disorder. Many patients develop intractable headaches, sixth nerve palsy, pulsatile tinnitus. There is also a risk of permanent and severe vision loss<sup>4</sup>. However in rare cases patient may be asymptomatic especially children.

IIH is defined by modified Dandy's criteria (table 1)

**Table 1**

Modified Dandy criteria
Symptoms of raised intracranial pressure
No localizing signs with the exception of abducens (sixth) nerve palsy
The patient is awake and alert
Normal CT/MTR findings without evidence of thrombosis
Lumbar puncture opening pressure of > 25 cm H <sub>2</sub> O and normal biochemical and cytological composition of Cerebrospinal Fluid (CSF)
No other explanation for the raised intracranial pressure

As per Durcan FJ et al<sup>1</sup> and Radhakrishnan K et al<sup>2</sup>, the total incidence of IIH in female between 15 to 44 years of the age group is 0.9/100,000 persons and 3.5/100,000. The incidence of IIH is as high as 19 per 100,000 in women who are obese and are between age group of 20 to 44 years.<sup>1</sup> More than 90% patients of IIH are women who are obese and are of childbearing age typically described as 3Fs i.e. fatty, female in forties. 10% may have recurrent symptoms and signs.

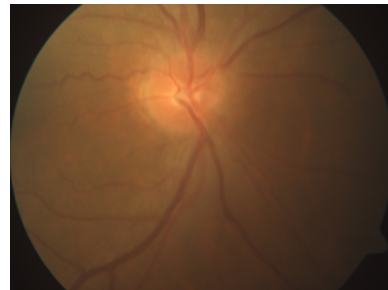
Treatment of IIH includes conservative management in form of avoiding the use of causative agents, reduction of weight and lowering of intracranial pressure with drugs like acetazolamide, furosemide etc. and surgical intervention like ventricular shunting or optic nerve sheath decompression and fenestration<sup>5</sup>

### CASE REPORT

A 65 years old male, having medium-built with (Body Mass Index = 24.0 kg/m<sup>2</sup>), presented for routine eye check up. He was a known hypertensive on tablet amlodipine 5mg OD for last 05 years. There was no history of headache, diminution of vision, transient obscuration of vision, double vision or tinnitus. No significant past and medication history.

On ocular examination, the best-corrected visual acuity was 6/6 with (-0.75 / -0.50 x 180°) and 6/6 with (-0.75/-0.50 x 180°) in right and left eye respectively. No relative afferent pupillary defect was seen. The

extraocular movements were full in both eyes without any pain and diplopia. Anterior segment examination was essentially normal in both eyes. Dilated examination of fundus revealed optic disc swelling associated with peripapillary haemorrhages (Figure 1a) in the left eye only. The right fundus was unremarkable (Figure 1b). The cup-to-disc ratio was 0.4:1 in right eye. Intraocular pressure was 22 mmHg and 14 mmHg in the right and left eye respectively with Goldmann applanation tonometry. Ishihara colour vision test was normal bilaterally. Humphrey visual field 30-2 SITA standard revealed an enlarged blind spot in the left eye only. Blood pressure was 142/88 mmHg right arm in supine position. Thorough investigations were done to rule out various causes of unilateral disc edema like AION, optic disc drusens, papillitis, foster kennedy syndrome and other infectious causes.



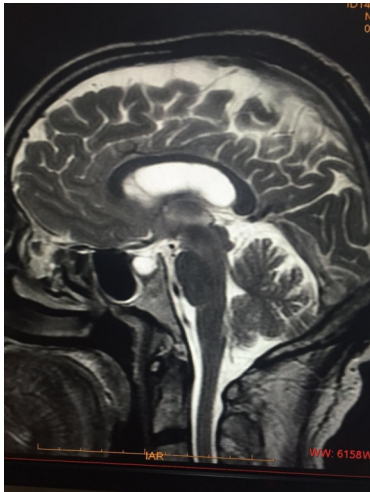
**Fig 1b: Normal optic disc (right eye)**



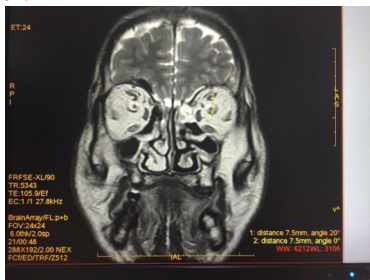
**Fig 1a: Optic disc swelling associated with haemorrhages suggestive of optic disc edema (left eye)**

In hematological investigations, complete blood count, ESR, fasting and postprandial blood sugar, HbA1C, thyroid functions test and viral markers were all normal. Carotid Doppler was normal for age. Contrast enhanced Magnetic Resonance Imaging (MRI) of the brain and orbits (Figure 2a) showed widened and partially empty sella without any features of sinus thrombosis and prominent CSF sleeve around the optic nerves bilaterally. Optic nerves measure 7.5 mm on both sides (Fig 2b). Consultation of neurophysician was taken wherein

lumbar puncture was done which showed a high opening CSF pressure of 268 mm of water, with normal CSF biochemistry contents. Thus on the basis of above reports, the diagnosis of Idiopathic intracranial hypertension was made.



**Fig 2a: Midline sagittal section of the brain showing empty sella filled with CSF.**



**Fig 2b: Showing prominent CSF sleeve around the optic nerves bilaterally. Optic nerves measure 7.5 mm on both sides.**

The patient was treated with oral tablet acetazolamide 500 mg twice a day. After 02 months the optic disc edema along with haemorrhages resolved leaving partial optic atrophy.

## DISCUSSION

There are various case reports on asymptomatic IIH in children which were diagnosed incidentally by bilateral disc edema<sup>3</sup>. Despite of raised intracranial pressure, reason for lack of headache in these children is still not known. Although these children may not have headache but they may have vision loss and neurological signs on presentation<sup>7</sup>. These children are treated in similar manner as patients who are symptomatic. Asymptomatic elderly male and unilateral disc oedema are the important features in our case and both are rare in case of IIH. Being very rare entity in elderly population, its incidence is not mentioned in the literature.

In our case there was unilateral presentation of papilledema. Various causes of unilateral optic disc edema like optic disc drusens, papillitis, AION etc. were ruled out. In view of the above red free fundus photography, ophthalmic ultrasonography and OCT was done to rule out optic disc drusens. In view of no RAPD at initial presentation and rare age group papillitis was ruled out. As patient has normal vision on initial presentation with no relative inferior altitudinal or absolute inferior nasal defect on perimetry, AION was ruled out at initial presentation. However Hayreh described a pre infarction state or incipient state of AION in which there is optic disc edema with enlarged blind spot and no visual field loss however this presentation is very rare and also in view of raised ICP this entity was unlikely thus after thorough investigation diagnosis of IIH was made.

Various hypothesis have tried to explain the occurrence of unilateral papilledema in IIH. It is thought to be dependent on various factors: optic disc perfusion pressure, intraocular pressure, CSF pressure.<sup>6</sup>

First, as per one hypothesis<sup>6</sup> there may be vascular disease sufficient enough to cause carotid or ophthalmic artery stenosis, and in turn responsible for the unilateral or asymmetric papilledema. However

carotid doppler was normal thus it was unlikely that there was any difference in optic disc perfusion pressure in the patient.

Second, there could also be a possibility that intraocular pressures between the two eyes could be asymmetric. In our case relatively higher intraocular pressure in the right eye (22 mmHg) as compared to left eye (14 mmHg), may lead the globe to withstand against greater scleral compression. However being relatively lower intraocular pressure in the left eye (14 mmHg), there is greater scleral compression due to the elevated intracranial pressure<sup>6</sup> and left eye is not able to withstand it as much as right eye thus giving rise to the disc swelling.

Third, there could be possibility of higher CSF pressure at the optic disc. It has been observed that in patients with unilateral or asymmetric papilledema, width of optic nerve sheath is increased and is usually symmetric.<sup>8</sup> In our patient despite of unilateral disc oedema we also found symmetrically enlarged optic nerve sheath (7.5 mm bilaterally) on MR imaging. Ultrasound (30° test) can also be used to aid in diagnosis of intracranial hypertension by measurement of the optic nerve sheath diameter. Multipurpose ultrasound units with high-frequency transducers (>7.5 MHz) can give high lateral and axial precision thus aiding in diagnosis.<sup>10</sup>

Hayreh<sup>9</sup> showed that the optic nerve sheath consists of fibrous tissue and after getting unfolded, it can expand till particular limit only. There may be differences among the patients in the trabecular meshwork of the optic nerve sheath. Whenever there is increased CSF pressure, the pressure in the optic nerve sheath may rise enough to distend the sheath, but not good enough at the optic nerve head to cause disc oedema.

## CONCLUSION

In cases of unilateral disc edema where patient is asymptomatic, apart from common causes like papillitis, optic disc drusens, AION, one should also have suspicion of this unique and unilateral presentation of IIH. Thorough investigations like lumbar puncture with CSF pressure measurement and neuro-imaging should be carried out to confirm this entity. Early management of this treatable condition can prevent permanent visual impairment.

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