

Unilateral Papilledema and Choroidal Folds: An Uncommon Presentation of Idiopathic Intracranial Hypertension

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Abstract

The typical presenting ocular signs of idiopathic intracranial hypertension (IIH) include bilateral papilledema, enlarged blind spots, and abducens nerve palsy. Unilateral papilledema with choroid folds is an atypical presenting sign of IIH. The authors report the case of a 55-year-old Chinese gentleman, of medium-built, presenting with right eye transient visual obscuration associated with tinnitus for 2 weeks. Physical examination revealed ipsilateral choroidal folds and disc swelling. Computerized tomography of the brain and orbit with contrast revealed no intracranial space occupying lesions. Lumbar puncture revealed an elevated opening cerebrospinal fluid pressure (30 cm H₂O) with normal biochemistry. Subsequent contrast Magnetic Resonance Imaging of the brain and orbit with venogram revealed features of empty sella, compatible with the diagnosis of IIH. He was subsequently treated with oral acetazolamide for 6 months and the symptoms resolved. Unilateral papilloedema with choroidal folds is an uncommon presentation of IIH, a lumbar puncture with opening pressure measurement and proper neuro-imaging should be considered when there is suspicion of IIH.

Keywords: Idiopathic intracranial hypertension; Papilloedema; Choroidal folds

Introduction

Idiopathic Intracranial Hypertension (IIH) or pseudotumor cerebri has also been previously termed benign intracranial hypertension. IIH is not as benign as it was once thought to be as many patients suffer from intractable and disabling headaches with a risk of permanent visual loss [1-3].

The annual incidence of IIH in the general population has been reported to be around 1-2 cases per 100,000, [4,5] with a higher prevalence among women aged 20-44 years (3.5 cases per 100,000) [6]. IIH is relatively uncommon in men, with a female to male ratio of approximately 4.3:1 to 8:1 [6,7].

IH should be distinguished from secondary intracranial hypertension produced by a neoplastic lesion. Diagnosis of IIH is based on the Modified Dandy criteria [8] (Table 1).

Typical presenting ocular signs of IIH include bilateral papilloedema, enlarged blind spots, and abducens nerve palsy [7,9]. There is a lack of randomized controlled trials investigating the optimal treatment for IIH and the natural history of untreated IIH is still relatively unknown. The primary aims of treatment in IIH are vision preservation and symptomatic control [4,9].

Treatment includes avoiding the causative agents, weight reduction, and intracranial pressure lowering with medications (acetazolamide, furosemide, painkillers, antidepressants or anticonvulsants) or surgeries (optic nerve sheath decompression and fenestration as well as ventricular shunting) [7,9,10].

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 ₂ O and normal biochemical and cytological composition of Cerebrospinal Fluid (CSF)
No other explanation for the raised intracranial pressure

Table 1: Diagnostic criteria of idiopathic intracranial hypertension

Case Report

A 53-year-old, medium-built (Body Mass Index = 24.5 kg/m²), retired fireman presented with on and off headaches and transient visual obscuration associated with pulsatile tinnitus for 3 months. He had hypertension but otherwise enjoyed good past health. The ophthalmic and medication histories were unremarkable.

On ocular examination, the best-corrected visual acuity was 6/9 (-1.00 / -0.75 x 180°) and 6/6 (-0.75/-0.50 x175°) over the right and left eye respectively. There was no relative afferent papillary defect. Anterior segment examination was normal with normal intraocular pressure (15 mmHg and 20 mmHg over the right and left eye respectively, measured with Goldmann applanation tonometry). The extraocular movements were full without any diplopia or pain. Dilated fundi examination revealed optic disc swelling with choroidal folds (Figure 1a) in the right eye only. The left fundus was unremarkable (Figure 1b). The cup-to-disc ratio was symmetrical and normal

bilaterally. There was no proptosis, lid retraction or lid lag. Ishihara color vision acuity test was full bilaterally.

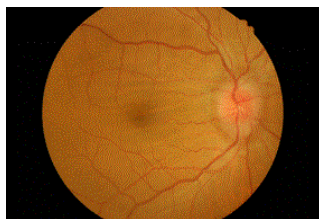


Figure 1a: Right eye choroidal fold with disc swelling



Figure 2b: Unremarkable left fundus

For the hematological investigations, complete blood count, thyroid function, and autoimmune marker were all normal. Fasting glucose was 5.0 mmol/L and the HbA1C was 5.9%. Blood pressure was 135/78 mmHg. Goldmann visual field revealed an enlarged blind spot in the right eye only (Figure 2).

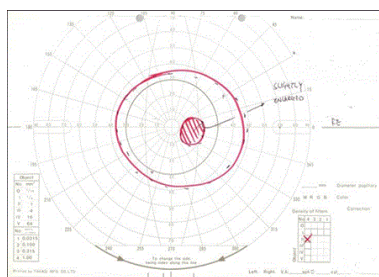


Figure 2: Right eye blind spot enlargement

Computerized tomography (CT) scan of the brain and orbits with contrast was normal without evidence of the “T-sign” nor posterior scleral thickening. There were no space occupying lesions detected (Figure 3).

Lumbar puncture in the lateral decubitus posture revealed a high opening CSF pressure of 30 cm H₂O, with normal CSF biochemistry contents.

Magnetic Resonance Imaging (MRI) of the brain and orbits with a venogram (Figure 4a and 4b) showed an empty sella without any features of sinus thrombosis. Based on the above features, the diagnosis of IIH was made.



Figure 3: Normal CT brain and orbits with contrast

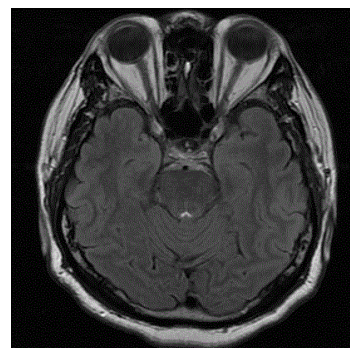


Figure 4a: T2 FLAIR MRI brain

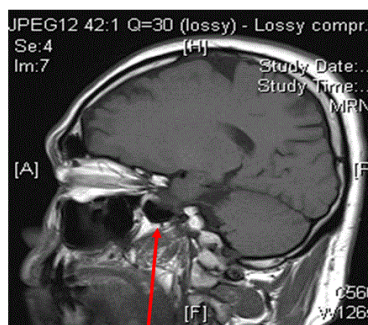


Figure 4b: Saggital MRI brain showing empty sella

The patient was treated with oral acetazolamide 500 mg twice a day for 6 months with gradually improvements in the headache and tinnitus and the transient visual obscurations also subsided. The right-sided papilloedem and choroidal folds only partially resolved on clinical examination at 1 year after completion of the treatment but the patient remained symptom free.

Discussion

Papilloedema is present in the majority of patients with IIH and usually occurs bilaterally [11], although unilateral occurrences were reported in 10% of cases [12,13]. Recent evidence revealed that

papilloedema is caused interruptions in the optic nerve (ON) metabolism as a result of the high intracranial pressure transmitted to the subarachnoid space surrounded the ON, leading to edema, ischemia, and eventual visual impairment or loss. Choroidal folds arise from globe compression from the distended optic nerve sheath leading to globe distortion and acquired hyperopic shift [15-21]. In our case, the patient reported no changes in his refractive errors compared to his pre-morbid state. In theory, choroidal folds may precede the formation of retrolaminar hypoperfusion and papilloedema [20].

The most interesting feature in this case is the asymmetrical presentation between the 2 eyes, which may be explained by the differences in intraocular pressure between the 2 eyes. The relatively higher intraocular pressure in the left eye (20 mmHg) may allow the globe to withstand greater scleral compression whilst the relatively lower intraocular pressure in the right eye (15 mmHg) may predispose it to greater scleral compression (choroidal folds) as a result of the elevated intracranial pressure swelling and the resultant retrolaminar hypoperfusion, giving rise to the disc swelling.

Unilateral acquired choroidal folds associated with papilloedema is a rare presentation of IIH. After ruling out space occupying lesions and other causing of unilateral optic neuropathy, clinicians should have a high index of suspicious of this unique and asymmetrical presentation of IIH. Further neuro-imaging and lumbar puncture with opening pressure measurement should be considered to prevent delay in the diagnosis and management of this treatable condition in order to prevent permanent visual impairment.

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