

Bilateral Buried Optic Nerve Head Drusen Mimicking Idiopathic Intracranial Hypertension

Juhi Sahu, Muthukrishnan Vallinayagam, C Alekhya

Department of Ophthalmology, Mahatma Gandhi Medical College & Research Institute, Pondicherry, Sri Balaji Vidyapeeth, India.

Abstract

Optic nerve head (ONH) drusen are buried deposits in the optic nerve head simulating papilloedema. Idiopathic Intracranial Hypertension (IIH) signifies raised intracranial pressure in the absence of intracranial space occupying lesion. A 35-year old woman diagnosed elsewhere with IIH and treated with oral acetazolamide for six months, presented with non-specific headache. Fundus showed bilateral blurring of disc margins, elevated disc and obscuration of cup. MRI of brain was normal. The non-resolution of disc edema despite six months of acetazolamide therapy raised suspicion of an alternate diagnosis. Ultrasound B-scan confirmed the diagnosis of ONH drusen. ONH drusen should be excluded before entertaining the diagnosis of IIH.

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Keywords: Buried Optic Nerve Head Drusen, Idiopathic Intracranial Hypertension, Pseudopapilloedema

Introduction

Optic nerve head (ONH) drusen are abnormal calcified mitochondrial deposits resulting from impaired axonal metabolism.¹ Idiopathic Intracranial Hypertension (IIH) is a headache syndrome characterized by papilloedema with normal cerebrospinal fluid (CSF) composition and normal neuroimaging.^{2,3} Pseudopapilledema is an apparent optic disc swelling secondary to a benign process such as buried Optic Nerve Head (ONH) drusen, hypoplastic, tilted or hyperopic disc. ONH drusen accounts for 75% of clinically diagnosed cases of pseudopapilledema.⁴

Case Report

A 35-year old woman diagnosed with Idiopathic Intracranial Hypertension (IIH) elsewhere and commenced on oral

acetazolamide therapy for six months, presented with intermittent episodes of "vague" headache. She denied ocular complaints. Barring the headache, there was no history suggestive of raised intracranial pressure. The visual acuity and pupillary examination were normal bilaterally. The fundus examination revealed bilateral blurring of disc margins, elevated disc and obscuration of physiological cup (Figure 1). MRI of brain was normal (Figure 2). Visual field charting (HFA 24-2) was unremarkable (Figure 3). The persistence of disc edema despite strict compliance with medical therapy for six months aroused a clinical suspicion with respect to an alternate diagnosis. Ultrasound B-scan showed highly reflective deposits which persisted on reducing gain, suggesting ONH drusen (Figure 4). The

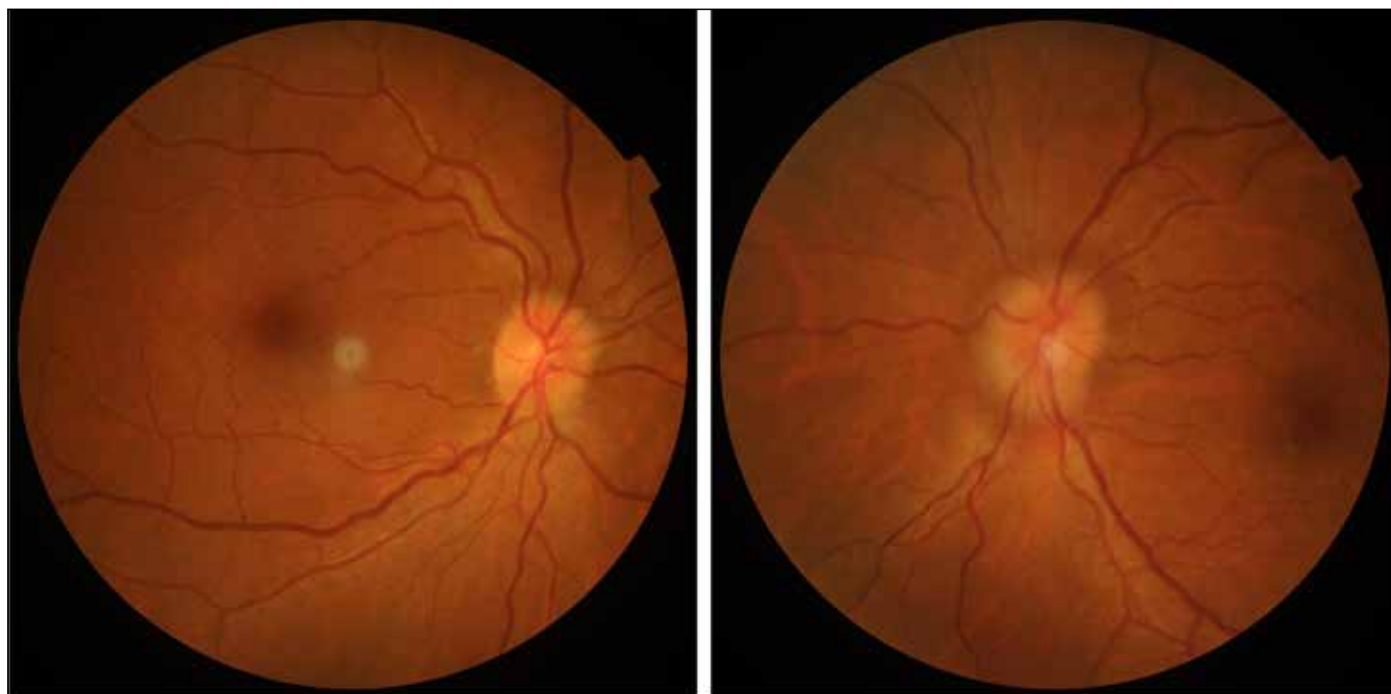


Figure 1: Fundus photograph showing bilateral blurring of disc margins, elevated disc and obscuration of physiological cup



Figure 2: MRI of brain showing normal picture

optic nerve sheath diameter was within normal limits. Acetazolamide therapy was discontinued and patient was reassured.

Discussion

Idiopathic Intracranial Hypertension (IIH) or pseudotumor cerebri is characterized by papilledema in the presence of normal cerebrospinal fluid composition and normal neuroimaging. Modified Dandy’s criteria for the diagnosis of IIH includes signs and symptoms of raised intracranial pressure (ICP), absence of localising findings on neurological examination, normal neuroimaging, elevated CSF opening pressure >200 mm of water (>250 in obese) in an awake and alert patient, with no other cause for raised ICP.

ONH drusen is a rare congenital optic disc anomaly seen in 2% population. It is hypothesized to occur due to abnormal optic disc vasculature causing axonal ischemia and impaired axonal metabolism. The optic disc appears elevated with indistinct margins and a localized lumpy appearance. ONH drusen tends to evolve over the years, from being buried to becoming exposed in the setting of optic atrophy and reduction of retinal nerve fiber layer (RNFL) thickness. Although often asymptomatic, ONH drusen is associated with visual field defects in 87% of cases which may be arcuate defects, enlargement of blind spot or peripheral constriction of visual field. Vision threatening complications like Anterior ischemic optic neuropathy (AION), retinal vascular occlusions, choroidal neovascular membrane and retinal haemorrhages have been reported.⁵ Though the classical fundus appearance confirms an exposed drusen, a definitive diagnosis of buried drusen remains a challenge.⁴

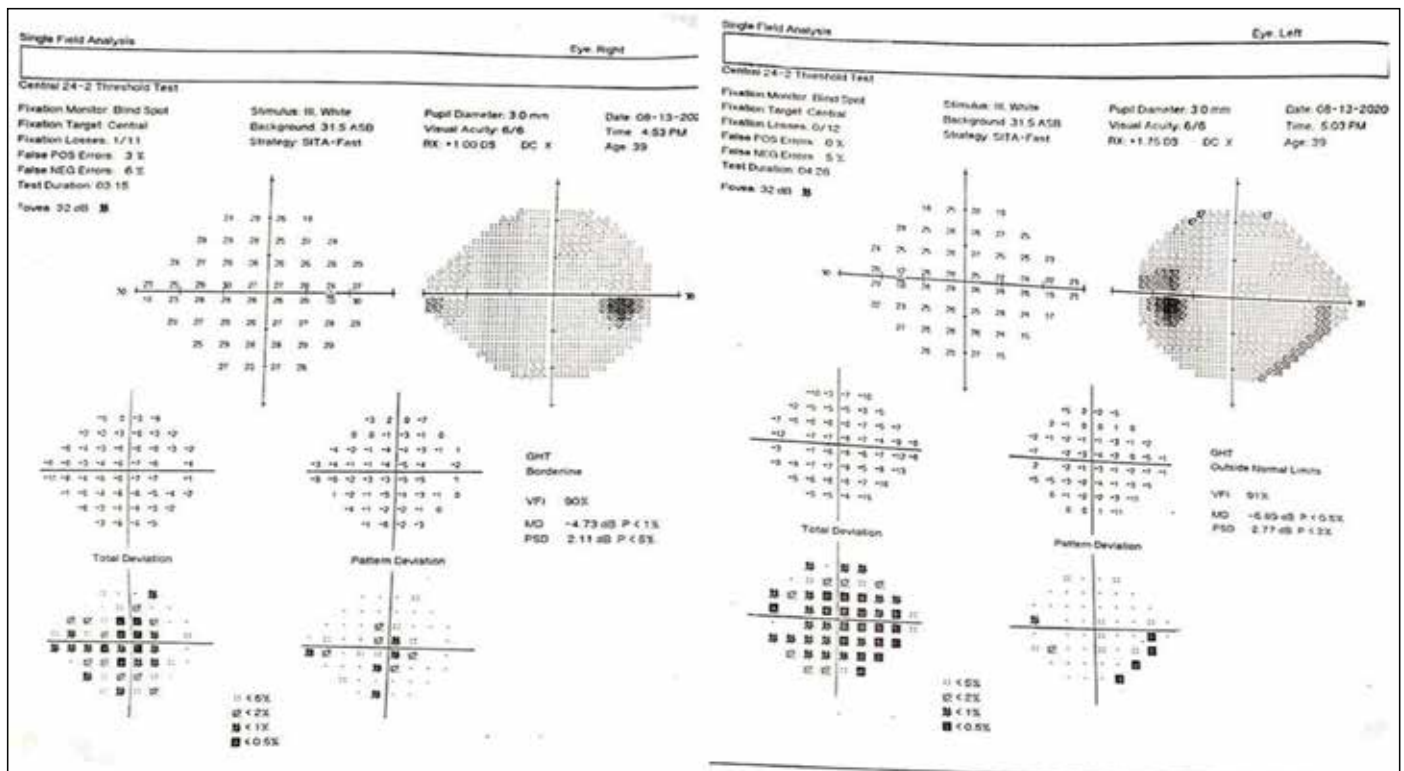


Figure 3: Visual field charting (HFA 24-2) was unremarkable

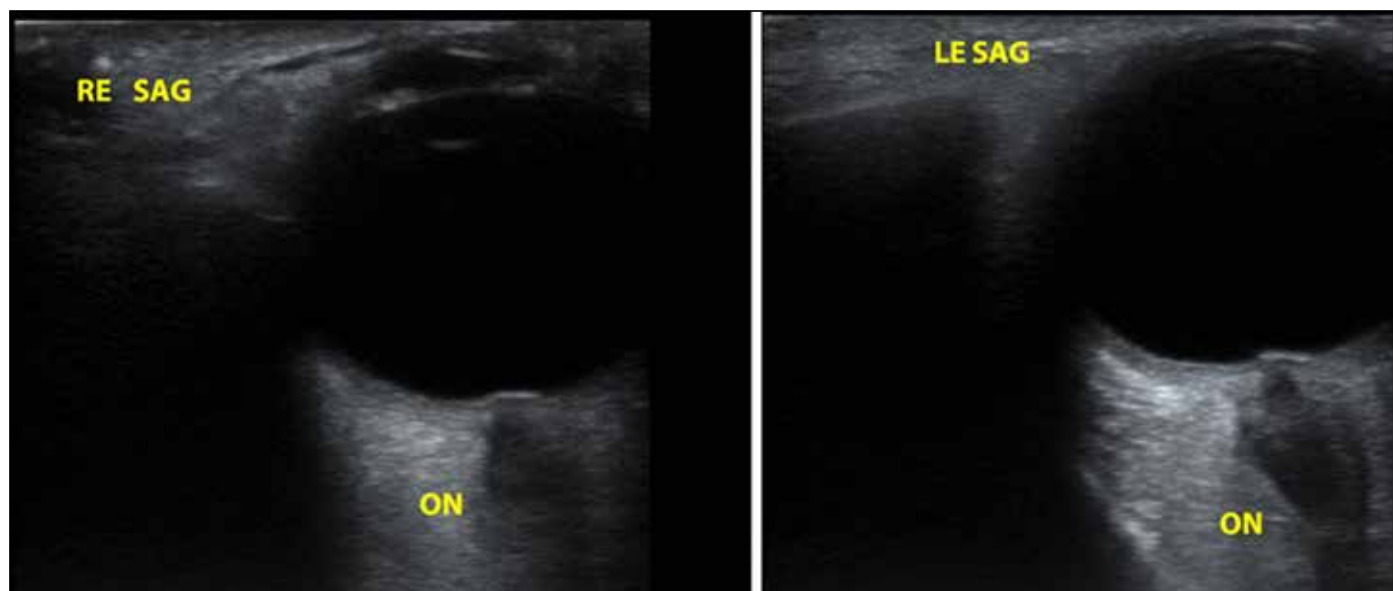


Figure 4: Ultrasound B-scan showed highly reflective deposits which persisted on reducing gain, suggesting ONH drusen

A buried ONH drusen can cause an elevated disc with blurred margins and thereby mimic papilloedema.⁵ Orbital ultrasound offers a non-invasive modality in establishing the diagnosis.

The diagnosis of IIH is to be reconsidered in a patient with an atypical clinical picture. The non-resolution of disc elevation following compliant medical therapy for IIH is unusual and hints towards an alternative diagnosis. ONH drusen as a differential for pseudopapilloedema should be excluded before initiating therapy for IIH.

Declaration Of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Address for correspondence

Muthukrishnan Vallinayagam
MS, DNB, FRCS, FICO, Associate Professor
Department of Ophthalmology,
Mahatma Gandhi Medical College
& Research Institute, Sri Balaji
Vidyapeeth, Pondicherry India.
Email: muthu2308@yahoo.co.in



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