Magnetic Resonance Imaging Findings in Idiopathic Intracranial Hypertension - 2 Cases with Review of Literature – A case report

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Abstracts: Idiopathic Intracranial Hypertension (IIH) also known as Benign Intracranial Hypertension and Pseudotumor cerebri is a headache syndrome of increased Intracranial Pressure (ICP) without ventriculomegaly or mass lesion and with normal Cerebrospinal Fluid (CSF) composition. Two cases of IIH who presented with headache, Magnetic Resonance Imaging (MRI) findings in the brain and orbits of these patients provided valuable information are presented herewith [Goswami G NJIRM 2014; 5(5):114-117]

Key Words: Idiopathic Intracranial Hypertension, MRI, Opening CSF Pressure

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Introduction: Idiopathic Intracranial Hypertension (IIH) also known as Benign Intracranial Hypertension and Pseudotumor cerebri is a headache syndrome of increased Intracranial Pressure (ICP) without ventriculomegaly or mass lesion and with normal Cerebrospinal Fluid (CSF) composition. Although this disease condition was described almost a century ago its pathogenesis remains unknown. Diagnostic and therapeutic developments in the past two decades have substantially improved the patient management.

We report two cases of IIH who presented with headache. Magnetic Resonance Imaging (MRI) findings in the brain and orbits of these patients provided valuable information regarding the diagnosis which was further confirmed by opening CSF pressure studies.

Case 1: 33 years old female patient presented to the Neurology OPD with complaints of non-resolving frontal headache occurring intermittently over several months. There was progressive worsening of her symptoms over the last two months. There was no associated vomiting, visual symptoms or any other complaint pertaining to the neurological system. On examination the patient was obese (BMI-31 kg/m^2). Her vital parameters were within normal limits (Pulse- 80/min, BP-116/82 mm Hg). Fundoscopic examination revealed bilateral papilloedema. Further ophthalomogical examination revealed a normal visual acuity and normal visual fields. There was no focal neurological deficit. Her laboratory parameters including routine blood and urine investigations were normal. Electroencephalographic (EEG) study was also normal.

MRI examination of the brain and orbits was performed using a 1.5 T Superconducting GE Signa system. On Fat Saturated T2W (FS T2W) images there was vertical tortuousity and elongation of optic nerves noted. The perioptic subarachnoid spaces were distended bilaterally with flattening of the posterior sclera of the globes.

MRI of the brain revealed small cortical veins and cerebral venous sinuses. No ventricular dilatation, intracranial mass lesion or sinus thrombosis was however indentified. Note was made of empty sella with inferiorly displaced and compressed pituitary gland.

Based on these findings a diagnosis of IIH was given. To further substantiate our findings a lumbar puncture with patient in left lateral position was performed. It revealed an opening pressure of 280 mm H_2O. CSF analysis revealed normal composition and there was no evidence of meningitis confirming our diagnosis of IIH.

The patient was put on weight management regime along with Carbonic Anhydrase Inhibitors and showed satisfactory progress during her stay in the hospital.

Case 2: A 19 year old female patient was referred for MRI of the Brain following complaints of throbbing frontal headache with periods of remission lasting six months. She complained of occasional blurring of vision associated with these

complaints. There was no history of fever, vomiting or any neurological deficit. On examination the patient was slender (BMI- 18 kg/m^2) and afebrile. Her vitals were stable (Pulse- 70/min, BP- 106/74 mm Hg). Her neurological examination did not reveal any cranial nerve or sensorimotor deficit. Fundoscopic examination showed bilateral papilloedema, however there was no disc atrophy. The routine laboratory investigations and EEG of the patient were within normal limits.

MRI examination of the orbits revealed elongated, tortuous optic nerves with prominent perioptic CSF spaces bilaterally on FS T2W images. Posterior scleral indentation was observed bilaterally. Study of the brain showed smaller caliber of the cortical veins and venous sinuses. No space occupying lesion, ventricular dilatation, evidence of meningitis or sinus thrombosis was seen.

Further confirmation of our working diagnosis of IIH was done by studying opening CSF pressure which was elevated (290 mm H_2O). CSF analysis was within normal limits.

The patient was managed on Carbonic Anhydrase Inhibitors and Corticosteroids and showed satisfactory improvement.

Case 1: Image 1: Axand Sag T2wi Images Show Small Cortical Veins And Cerebral Venous Sinuses With No Ventricular Dilatation, Intracranial Mass Lesion Or Sinus Thrombosis. Empty Sella with Inferiorly Displaced and Compressed Pituitary Gland

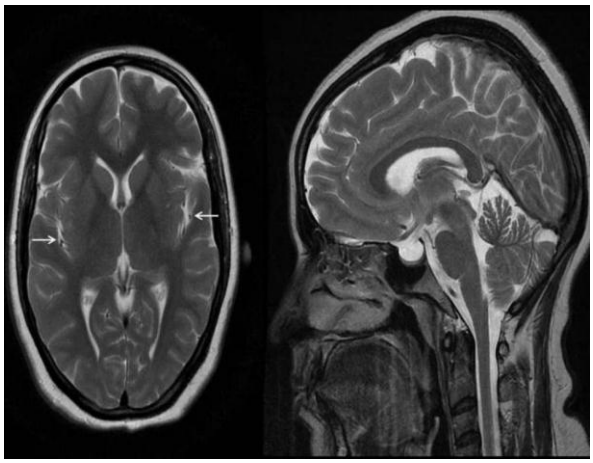
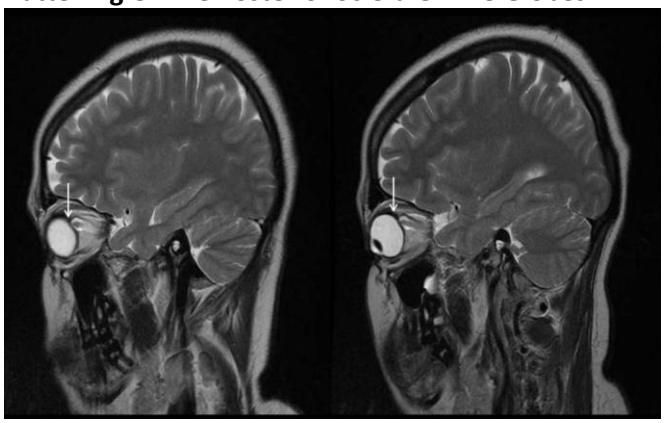


Image 2: Sag FS T2WI Show Vertical Tortuousity and Elongation of Optic Nerves. Distended Perioptic Subarachnoid Spaces Bilaterally With Flattening Of The Posterior Sclera Of The Globes



Case 2: Image 1: AX T2WI Image Shows Smaller Caliber Of The Cortical Veins And Venous Sinuses On T2WI

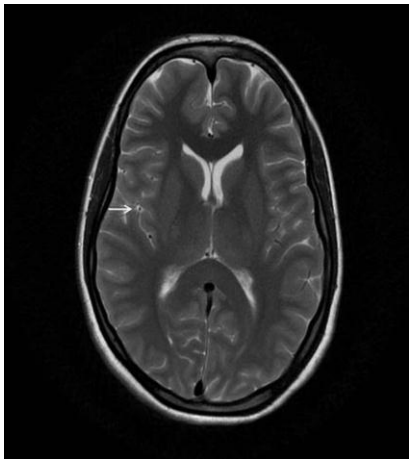
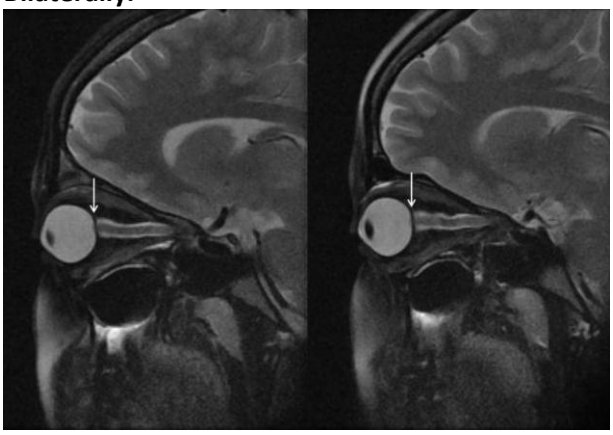


Image 2: Sag Fs T2wi Prominent Perioptic Csf Spaces With Posterior Sclera Indentation Bilaterally.



Discussion: IIH is a syndrome characterized by headache, increased ICP and papilloedema in

patients without focal neurological findings except for occasional VI cranial nerve palsy. It is common in young overweight women with an annual incidence as high as 20 per 100,000 in this subgroup. Currently the following criteria are assessed for diagnosis of IIH- a) symptoms and signs attributable to increased ICP or papilloedema b) elevated ICP recorded during the lumbar puncture c) normal CSF composition d) no imaging evidence of ventriculomegaly or a structural cause for increased ICP e) no other cause of intracranial hypertension identified inclusive of certain medications [(1)].

A diagnosis of IIH is therefore based on assessment by imaging, CSF opening pressure studies, visual field testing, monitoring of the optic nerve head, visual acuity testing, contrast sensitivity and visual evoked potentials [1]. An opening CSF pressure of greater than 250 mm H_2O with patient in left lateral position is consistent with diagnosis. Pressures less than 200 mm H_2O are normal and those between 201-249 mm H_2O are non-diagnostic[2].

Traditionally imaging of the brain has been performed in these cases to rule out any secondary cause for raised ICP but now MRI examination of the brain and orbits is being used more frequently to study this abnormality. Jinkins et al[3] and Gass et al[4] reported distension of the perioptic subarachnoid space and ballooning of optic papilla secondary to elevated ICP which causes it to protrude physically into the posterior aspect of globe.

In long standing cases of raised ICP a downward herniation of the suprasellar CSF space through a defect in diaphragma sella was noted in resulting in classic empty sella [5, 6].

Brodsky et al reported flattening of posterior sclera in 80%, an empty sella in 70%, distension of perioptic subarachnoid space in 45%, enhancement of prelaminar optic nerve in 50%, vertical tortuousity of the orbital nerve in 40% and intraocular protrusion of prelaminar optic nerve in 30% of 20 patients of IIH studied by MRI examination of the brain and orbits [7].

Soler et al[8] reported that four of 22 paediatric patients with IIH showed increased ICP in the absence of papilloedema. Similar findings were endorsed by Suzuki et al [9] in their case report.

Zagardo et al [5] and Suzuki et al [9] repeated MRI in patients with IIH and found that optic nerve and pituitary abnormalities returned to normal after normalization of the CSF pressure. It is therefore imperative that patients with severe headache be evaluated with MRI examination of the brain and orbits to document such cases of raised ICP without papilloedema. In our study of two cases the optic nerve abnormalities were well documented. In addition empty sella was seen in one case. These findings further undermine the importance of MR imaging of the brain and orbits not only to rule out the lesions that produce raised ICP but also to make a primary diagnosis of IIH in the presence of optic nerve and pituitary abnormalities. Follow up MRI for evaluation of treatment response is recommended with demonstration of return to normal appearances on normalization of the ICP.

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