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MAGNETIC RESONANCE IMAGING FINDINGS IN IDIOPATHIC INTRACRANIAL HYPERTENSION

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ABSTRACT

Background and Purpose: Idiopathic intracranial hypertension (IIH) can alter the configuration of anatomic structures of the brain and can be associated with abnormal MRI findings. We aimed to assess the MRI signs suggesting elevated intracranial pressure (ICP) that are found in patients with IIH.
Materials and Methods: 46 Patients with IIH were investigated with Magnetic Resonance Imaging (MRI) and Magnetic Resonance venography (MRV) studies which were evaluated by two neuroradiologist to assess the presence or absence of transverse sinus stenosis (TSS), posterior globe flattening, optic nerve sheath dilation/tortuosity, and the size/appearance of the sella turcica.
Results: 42 patients with mean age 26 years were included, of them; (66.6%) had TSS, partial empty sella was found in (69%), Pituitary displacement in (33.3%), tortuous optic nerve in (61.9%), flat posterior sclera were found in (59.5%), Optic nerve sheath distension in (52.4%), and Optic nerve protrusion in (26.2%) of cases.
Conclusions: The presence of the previous MRI abnormalities helps to increase the diagnostic certainty of IIH. In atypical clinical situations (in absence of papilloedema or normal CSF opening pressure), the presence of any combination of these findings may necessitate further clinical evaluation.

INTRODUCTION

Idiopathic intracranial hypertension (IIH), also known as pseudotumour cerebri, is characterized by increased cerebrospinal fluid (CSF) pressure in the absence of an identifiable structural cause (Owler *et al*., 2005). IIH most commonly presents with manifestations of raised intracranial pressure, including headache, visual disturbances including transient visual obscuration, pulsatile tinnitus, and papilloedema in women with obesity of childbearing age (Friedman *et al*., 2002). IIH is not always benign; severe irreversible visual loss is possible (Hoffmann *et al*., 2013). Diagnosis is typically confirmed by a lumbar puncture, which demonstrates raised CSF pressure with normal composition. Magnetic resonance imaging (MRI) has been traditionally used to rule out other causes of increased intracranial pressure, mainly, intracranial space occupying lesions, hydrocephalus, or dural sinus thrombosis. Recently certain MRI findings have been reported to be associated with IIH, including flattening of the posterior wall of the sclera, protrusion of the intraocular portion of the optic nerve, vertical tortuosity of the optic nerve, optic nerve sheaths distension, enhancement of the optic nerve head and partial empty sella turcica (Hoffmann *et al*., 2013). As regard to magnetic resonance venography (MRV), a high proportion of patients with IIH have been found to have unilateral or bilateral transverse sinus stenosis. However, these imaging findings are non-specific, the combination of findings, in addition to the presence of non-thrombotic transverse sinus stenosis on MRV, would be expected to increase the diagnostic certainty for IIH (Hoffmann *et al*., 20013 and Ahmed *et al*., 2001). The objective of our study was to delineate the MRI findings in patients with clinically definite IIH. We hypothesized that if these presumed signs of elevated ICP are consistently present

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in all patients, this could further aid the diagnosis of clinically doubtful cases especially those without papilledema or normal CSF opening pressure. Also, the controversy of sinus stenosis was addressed.

MARETIALS AND METHODS

We retrospectively reviewed medical records of patients who was diagnosed clinically as IIH, and underwent standardized contrast enhanced brain MRI and contrast enhanced MRV from January 2014 to December 2014 at IbnSina Hospital, Kuwait, which is a tertiary hospital. We included all patients with primary IIH (those who found to have cerebrovenous thrombosis (CVT) were excluded). The diagnosis of IIH was made on the basis of the updated Modified Dandy criteria (Friedman *et al*.,2002 and Friedman *et al.,* 2013), papilledema, symptoms of raised ICP, no other cause for raised ICP on brain MRI, no CVT on brain MRV, and normal CSF contents with elevated CSF opening pressure (≥25 cm of water). Demographic information was recorded, including age, gender and body mass index (BMI). Results of lumbar puncture, especially CSF opening pressure, were documented. Only patients who had undergone MRI and MRV as part of their diagnostic work up were included in the study.

The scans of all identified cases were reviewed by two different neuroradiologists to rule out acute thrombosis on the static images, to look for abnormalities of flow on MRV and other signs on cross-sectional routine MRI study. The results of the neuroradiologists were grouped for each sign as 'sign present' or 'not present' for all of the following signs: transverse sinus stenosis (TSS), partially empty sella, posterior displacement of the pituitary stalk, flattening of the posterior wall of the sclera, intraocular protrusion of the optic nerve, distension of the optic nerve sheath and vertical tortuosity of the optic nerve. The study was approved by our Institutional Review Board.

RESULTS

Among the cases diagnosed with IIH, 46 patients underwent standardized contrast enhanced brain MRI and contrast enhanced MRV, 4 patients were excluded due to the presence of CVT. 42 patients included, of which 41 females and 1 male, their age ranged from 18 to 47 (26±2.3), BMI ranged from 22 to 35 (27.4±3.6) and the CSF opening pressure ranged from 290 mm water to 550 mm water (320±17.2). The demographic data was summarized in Table (1).

Table 1. Demographic data of IIH cases (N= 42)

Age	26±2.3(Mean ±SD)
Sex	
Female	41
Male	1
BMI	27.4±3.6
CSF pressure (in mm water)	320±17.2

The radiological findings in our patients were: transverse sinus stenosis (TSS) in 28 patients (Figure 1), partially empty sella was present in 29 patients (Figure 2), posterior displacement of the pituitary stalk in 14, tortuosity of the optic nerve in 26 patients (Figure 3), flattening of the posterior globe in 25 patients (Figure 4), optic nerve sheath distension in 22 patients (Figure 4 &5), and optic nerve protrusion in 11 patients and. These results are summarized in Table (2).

Table 2. Radiological findings of cases (N= 42)

Abnormality	N (%)
TSS	28 (66.6%)
Partial empty sella	29 (69%)
Pituitary displacement	14 (33.3%)
Tortuous optic nerve	26 (61.9%)
Flat posterior sclera	25 (59.5%)
Optic nerve sheath distension	22 (52.4%)
Optic nerve protrusion	11 (26.2%)

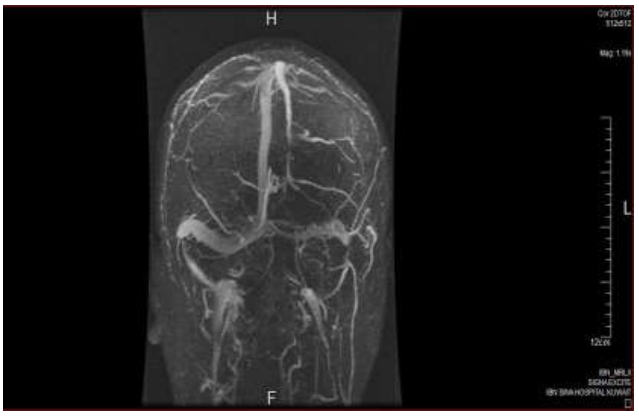

Figure 1. Hypoplastic left transverse and sigmoid sinuses

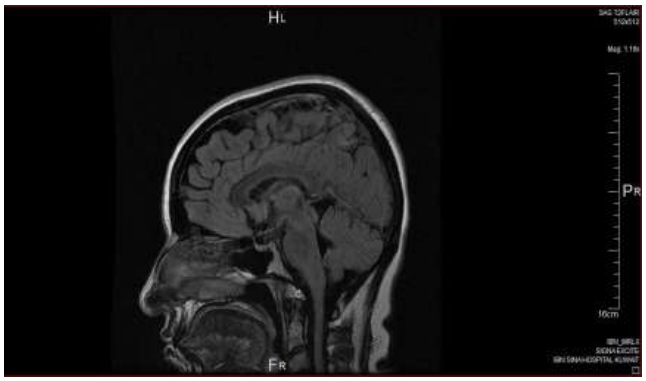

Figure 2. Partial empty sella

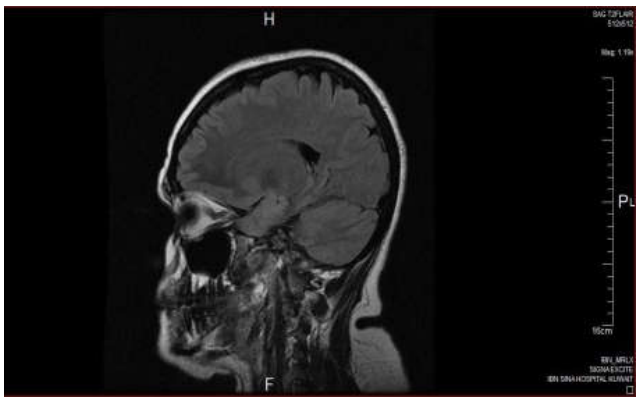

Figure 3. Tortuous optic nerve

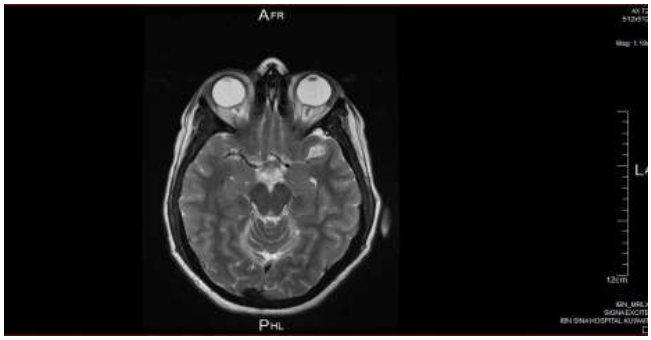

Figure 4. Bilateral hydrops and scleral flatness

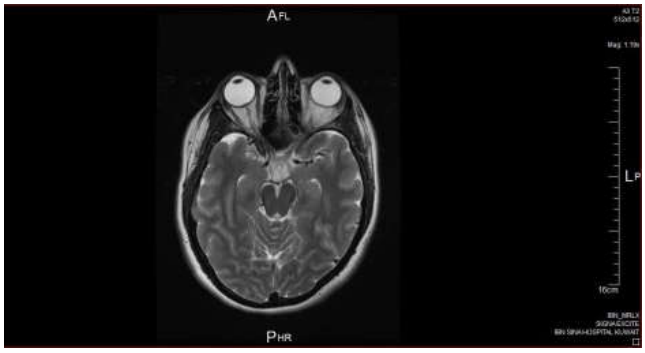

Figure 5. Optic nerve hydrops

DISCUSION

In a patient with the appropriate clinical phenotype, the diagnosis of IIH is essentially one of exclusion of other secondary causes of raised ICP (Sussman *et al*., 1998). MRI will identify any intracranial mass or hydrocephalus, MRV and MRI will identify venous sinus thrombosis. Acute thrombosis is not usually difficult to diagnose on the static images and will be confirmed by abnormalities of flow on MRV. In this respect, MRV of the superior sagittal sinus does not normally present any problems in interpretation, a normal study generally giving uniform signal over the course of the sinus (Sylaja *et al*., 2003). The lateral sinuses, however, are more difficult to assess, owing to a wide variation in the normal anatomy including, dominance or absence of either side, and they contain filling defects frequently (Ayanzen *et al*., 2000). In our study we found TSS in 66.6% of patients, in accordance with Higgins *et al* who found that the great majority of patients with IIH had a distinctive pattern on MRV signal gaps in both lateral sinuses, which was not seen in their ‘‘supernormal’’ control group (whom only exceptionally ever suffered from headache) (Higgins *et al*., 2004).

A TSS is found on MRV in most patients with IIH, with a specificity of 93% and a sensitivity of 93%. The stenosis can be “a smooth, tapered narrowing” due to extrinsic compression or “a discrete intraluminal filling defect” causing intrinsic obstruction or a combination of both (Farb *et al*., 2003). It was proposed on the basis of mathematic models that the collapsible transverse sinus might be compressed by ICH causing venous outflow obstruction, which results in further venous hypertension, then decreases CSF absorption and causes further increases in ICP, which then feeds back causing further external compression of the transverse sinus and further stenosis (Stevens *et al*., 2007). On the other hand, lowering the ICP can result in normalization of venous morphology, suggesting that this stenosis might be induced by IIH itself (Rohr *et al*., 2007). So yet controversy remains as to whether venous sinus stenosis is secondary to or is causative of the raised ICP (Strydom *et al*., 2010). This means that, regardless of cause or effect, many patients with IIH have venous hypertension and stenosis of either the dominant or both transverse sinuses (Owler *et al*., 2005), and it should not be mistaken for dural sinus thrombosis (Ayanzen *et al*., 2000). Apart from MRV changes, cross-sectional MRI signs of IH have been studied in patients with IIH with varying results and it was stated clearly that prediction of IIH may be possible by the trained neuroradiologist (Yuh *et al*., 2000). MRI features of IIH probably do not influence patient treatment, but they may be of use on follow-up, by using follow-up MRI it was indicated that MRI signs of IIH are at least partially reversible on normalization of ICP (Rohr *et al*., 2011 and Degnan *et al*., 2011).

Soler *et al* reported that four of 22 IIH patients showed increased CSF pressure in the absence of papilledema. They stated that attention to the optic nerves and pituitary gland should be given to MRI of patients with severe headache so as not to miss IIH without papilledema (Soler *et al*., 1998). Therefore, Normal optic discs are insufficient to exclude the presence of ICP. Moreover, Because of the natural fluctuations of ICP in IIH, measuring opening pressure does not always give the true steady-state pressure in otherwise appropriate clinical settings (Johnston *et al*., 1991), in patients with IH, including IIH, abnormal ICP waveforms are present. Although an elevated steady-state ICP has been reported in up to 93% of IIH patients (Gjerris *et al*., 1985), many patients show long periods of low or even normal ICP between short periods of marked intracranial hypertension (Gjerris *et al*., 1985 and Ball *et al*., 2006). Therefore increases in ICP may be missed by a single measurement and repeated lumbar puncture may be necessary if the clinical situation is highly suggestive of increased ICP (Friedman *et al*., 2002 and Stevens *et al*., 2008). Consistent with prior studies, seven MRI signs were found to be more prevalent in IIH patients; partially empty sella, pituitary stalk posterior displacement, flat posterior sclera, optic nerve protrusion, optic nerve sheath distension, optic nerve vertical tortuosity, and transverse sinus stenosis.

In the present study, our IIH patients actually exhibited partially empty sella in (69%), pituitary gland displacement in (33.3%), vertical tortuosity of the optic nerve (61.9%), flattening of the posterior sclera in (59.5%), distension of the perioptic subarachnoid space (52.4%) and optic nerve protrusion in (26.2%) of patients on initial MRI. Brodsky *et al* and Horoko *et al* recognized the MRI signs in their IIH patients were flattening of the posterior sclera in (80%), partially empty sella in (70%), distension of the periobtic subarachnoid space in (45%), tortuosity of the optic nerve in (40%) and enhancement of the optic disc in (50%). Slit-like ventricles on neuroimaging are neither sensitive nor specific for IIH (Brodsky*et al*.,1998 and Suzuki *et al*., 2001). Partially empty sella was detected in (69%) of our patients. The “empty sella” sign is associated with the long standing effects of increased ICP and is thought to result from a downward herniation of an arachnocele through the diaphragm sella

(Silbergleit *et al*., 1989). Wide range of sensitivities and specificities reported, in one study sensitivity was 65% while specificity was 95% (Agid *et al*., 2006) and in another it was 95 and 70% respectively (Maralani *et al*., 2012). Flattening of the posterior sclera was found in (59.5%) of our patients. Posterior globe flattening is considered by some authors to be the sine qua non neuroimaging sign of IIH and can be seen on both CT and MRI, but may be a more subtle finding subject to interpretation (Agid *et al*., 2006). Globe flattening may be explained by the transmission of elevated CSF pressure through the subarachnoid space, extending through the optic nerve sheath to the posterior globe (Maralani *et al*., 2012).It was stated that this sign is likely to be indicative of intracranial hypertension (Brodsky *et al* 2004 and Sajjadi *et al*., 2006), with asensitivity of 53.5% and a specificity of 100% (Maralani *et al*., 2012).

Intraocular protrusion of the optic nerve head was found in 26.2% of our patients is thought to occur in a manner similar to posterior globe flattening and is another sign associated with IIH (Grass *et al*., 1996 and Jinkins *et al*., 1996). Sensitivity was 37.2% while specificity was 100% (Maralani *et al*., 2012). Optic nerve tortuosity and optic nerve sheath distension were also associated with increased IIH; the distal and proximal points of fixation of the optic nerve enable it to kink freely in its course to the globe on protrusion of the intracranial contents under pressure (Brodsky,1998 and Grass *et al*., 1996). In our study optic nerve tortuosity found in 61.9% and sheath distension in 52.4%. Agid*et al* reported the sensitivity and specificity of these signs (40% and 91.1%) and (66.7% and 82.1%) respectively (Agid *et al*., 2006).

Conclusion

The presence of TSS, posterior globe flattening, optic nerve sheath dilation/tortuosity, and the size/appearance of the sellaturcica in brain MRI and MRV helps to increase the diagnostic certainty of IIH. In atypical clinical situations (in absence of papilloedema or normal CSF opening pressure), the presence of any combination of these findings may necessitate further clinical evaluation.

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The authors report no conflicts of interest.

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