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Value of double - track sign in differentiating primary from thrombosed transverse sinus stenosis in patients presumed to have idiopathic intracranial hypertension

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ABSTRACT

Background: Idiopathic intracranial hypertension (IIH) is primarily a disorder of young obese women of unknown etiology. The clinical presentation of IIH is similar to that of sinus thrombosis. The incidence of transverse sinus stenosis (TSS) reaches up to 90% of patients with IIH compared with normal subjects, and venous sinus thrombosis was reported in 11.4% of patients previously diagnosed as having IIH. Patients with thrombosis showed an abnormal region of double- track pattern on gadolinium (Gd) – enhanced T1WI within the dural sinus.

Aim: This study aims to evaluate whether double - track sign can differentiate primary TSS from thrombosed TSS in patients presumed to have IIH based on Gd - enhanced MRI.

Methods: This study was a retrospective multicenter observational case control study. The clinical and radiological data for all adult patients with presumed IIH were collected. The diagnosis of TSS was made based on further evaluation by DSA or MRV.

Results: Fifty-nine sinuses were diagnosed as a transverse sinus stenosis. Eight sinuses (13.6%) were partially occluded by recanalized thrombus. Double track sign was detected in seven (87.5%) of the thrombosed sinuses.

Conclusion: The double track sign remains much sensitive for the detection of transverse sinus thrombosis (TST) and it might provide an early clue for the dural sinus thrombosis in patients presumed to have IIH.

1. Introduction

Idiopathic intracranial hypertension (IIH) is basically a disorder of anonymous etiology that affect young obese women (< 45 years old), yet it can occur at any age, and less commonly affect adult males. Its overall annual incidence among general population is about two per 100,000 [1]. Approximately a quarter of patients will lose their vision from papilledema [2]. The clinical presentation of IIH resembles that of sinus thrombosis, this prompts the proposal that IIH is brought about by venous hypertension. However, rather than thrombus interfering with venous blood flow, venous hypertension is thought to occur as a result of stenosis of the transverse sinus specially its distal segments [3]. The frequency of transverse sinus stenosis (TSS) is very high in patients with IIH in contrast to normal subjects [4–6], and venous sinus thrombosis was confirmed in 11.4% of patients already diagnosed as having IIH [7].

Digital subtraction angiography (DSA) is the most sensitive investigation for detection of dural sinus lesions; however, the American Heart Association, and American Stroke Association recommend magnetic resonance imaging (MRI), and magnetic resonance venography (MRV) as the investigation of choice for the recognition of dural sinus lesions [8]. Chronic or partially recanalized thromboses are not well recognized on MRI, but may be obvious on MRV, usually as an extensive areas of narrowing or flow gaps [9] that sometimes resemble primary sinus stenosis [4]. On the other hand, MRV alone may have false-positive impression in cases of sinus aplasia, or hypoplasia (seen as a flow gap). It can also mistake T2-weighted hypointense signal of deoxyhemoglobin and intracellular methemoglobin as flow void [10].

It was observed that patients with thrombosis showed an abnormal region of double-track pattern on gadolinium (Gd) – enhanced T1WI within the dural sinus. The double track sign presented with two hyperintense linear tracts of contrast agent separated by the hypointense signal of non-contrast thrombus within dural sinus [11].

2. Aim

The aim of our study was to evaluate whether double - track sign can differentiate primary TSS from thrombosed (secondary) TSS in

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patients presumed to have IIH based on Gd - enhanced MRI.

3. Subjects and methods

This study was a retrospective multicenter observational case control study. Cases were recruited across 3 participating Hospitals in Saudi Arabia from January 2013 to January 2017. The research methodology was approved by ethics committee of College of Medicine, Prince Sattam bin Abdulaziz University.

We arranged with the neurologist and the radiologist to collect the clinical and radiological data for all adult patients (age > 18 years) with presumed IIH based on clinical features including: - symptoms and or signs (if present) that reflect generalized intracranial hypertension or papilledema, elevated CSF opening pressure, normal CSF composition, and normal CT scan of the brain [12]. The diagnosis of TSS was based on further evaluation by DSA or MRV, and correlated with the clinical presentation. The duration between MRV or DSA, and MRI was < 6 h. Patients without available images or unclear images were excluded from the study.

Adult patients with normal transverse sinus based on MRV or DSA, whether or not they fulfill the diagnostic criteria of IIH, were used as a control group. They were selected from the medical record of the participating centers.

3.1. Image interpretation

MR imaging was performed on 1.5 T scanners (Magnetom Vision or Symphony; Siemens, Erlangen, Germany). The radiologist at each hospital was responsible for data collection and the initial imaging assessment for primary or secondary TSS on DSA or MRV, then all the data and final imaging assessment were evaluated by the author. Each image was assessed independently on the clinical data. The reader evaluated the axial Gd-enhanced T1WI for the presence or absence of the double-track sign, and whether its presence indicates the diagnosis of secondary TSS or not.

After evaluating all images, reader collaboratively reviewed the clinical records to take the clinical and imaging information into account. He then determined the presence of primary or secondary TSS based on the results of MRV or DSA.

3.2. Statistical analysis

Analysis of data was done by IBM computer using SPSS (statistical program for social science) (version 10). Categorical variables were reported as number and percent. Continuous variable was presented as mean and standard deviation.

4. Results

In the present study; 82 patients who were presumed to have IIH, based on clinical and neuroimaging finding, were recruited over a 4-year period. Forty-three patients were excluded because of insufficient clinical, or imaging data, identification of a cause of intracranial hypertension other than transverse sinus thrombosis, or age ≤ 18 years. Thirty-nine patients were included and subsequently evaluated, 25 (64.1%) of them were female. The mean (± SD) age was 38.3 ± 11.2 years. Twenty-four patients with normal transverse sinus, 15 (62.5%) of them were female, with a mean (± SD) age of 38.1 ± 11.9 years, were included as a control group. So, 48 normal transverse sinuses were included as a control sinus.

All of our patients were evaluated with MRV, but 2 were evaluated by DSA. According to DSA or MRV; none of our patients had transverse sinus occlusion. Thirty-three patients (84.6%) had TSS, of them 26 (78.8%) had bilateral TSS (Fig. 1), and 7 (21.2%) had unilateral TSS. Therefore 59 sinuses were diagnosed as a transverse sinus stenosis. Eight (13.6%) of the stenosed sinuses were partially occluded by recanalized thrombus. The double track sign (Fig. 2) was detected on axial Gd-enhanced MRI in 7 (87.5%) of the thrombosed sinuses (6 cases of the unilateral TSS). No double track sings documented in the normal sinuses.

5. Discussion

Idiopathic intracranial hypertension (IIH) is characterized by headache, often on a daily basis, papilledema, transient visual obscurations, diplopia, vertigo and tinnitus The clinical picture is associated with abnormally high CSF pressure, normal CSF composition, and normal neuroimaging results, in a patient who is awake and alert without localizing signs on neurological examination [13].

The exact etiology of IIH is still unknown. However, various conditions may be associated with this syndrome. These include drugs (minocycline and tetracycline, growth hormone, steroids and vitamin A) [14], oral contraceptives use [15], hyperthyroidism [16], hypothyroidism [17], testosterone deficiency [18], acquired or congenital prothrombotic states [19], systemic lupus erythematosus [20,21], uraemia [22], obstructive sleep apnoea syndrome [23], extracranial venous hypertension [24], and iron deficiency anaemia [25].

Venous sinus stenosis has been identified in large majority of patients with IIH [4,5]. Bilateral transverse sinus stenosis is reported in 90% of patients with IIH, while, unilateral stenosis is reported in 9.8% [6]. This is in accordance with our results in which > 80% of our patients had TSS; and > 75% of them had bilateral TSS. It is not clear whether TSS is the result or the cause of increased intracranial pressure. Cerebral venous sinus stenosis will create a state of venous hypertension that will interfere with CSF absorption through the arachnoid granulations. As a result of that the intracranial pressure will be increased, which in turn will lead to further cerebral venous compression [26,27]. On the other hand, other studies suggest that venous flow disturbances in IIH are most probably the effect of CSF hypertension and not the cause [28–30].

We found that 13.6% of our patients with TSS have recanalized transverse sinus thrombosis. In the study of Agarwal et al., 11.4% of patients with presumed IIH had cerebral venous sinus thrombosis (CVST) after MRI and MRV evaluation; the most commonly affected sinuses were the superior sagittal sinuses, transverse and sigmoid sinuses [7]. It is important to recognize, and differentiate primary TSS from transverse sinus thrombosis (TST), that accounts for about 40% of all CVST [31], in patients with presumed IIH because the management protocols are entirely different.

In the present study, double track sign is detected on axial Gd-enhanced MRI in 87.5% of the thrombosed sinuses, and the double track sign is not documented in the normal sinuses. The double track sign presented with two hyperintense linear tracts of contrast agent, separated by the hypointense signal of non-contrast thrombus within dural sinus [11]. The double track sign is thought to result from the interaction between the damaged cellular wall and the thrombosis, the injury of endothelial cells with associated increased permeability of sinus wall [11]. CT is sensitive to detect acute dural sinus thrombosis [32], but it has no role in the chronic stage of thrombosis [33]. In addition, unenhanced MRI T1WI is unreliable for the detection of dural venous thrombosis [34]. In the study of, the signal intensity of thrombus on T1WI and T2WI was changed with the age of thrombosis, while, the signal intensity of the double-track sign on axial Gd-enhanced T1WI persisted throughout the different stages of TS thrombosis (acute, subacute and chronic), and it disappeared only in totally occluded or totally recanalized sinus, accordingly, the double-track sign usually indicate the TST [11]. For this reason; the double-track sign is considered as a direct sign in the identification of TST.

In conclusion, because the CT is sensitive for detection of acute TST, and the chronic thromboses or partially recanalized thromboses are less obvious on MRI, at the same time MRV alone may give a false-positive impression, the double track sign remains much sensitive for the