Clinical profile of cerebral venous sinus thrombosis and the role of imaging in its diagnosis in patients with presumed idiopathic intracranial hypertension

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Retrospective descriptive study reporting the rate of occurrence of cerebral venous sinus thrombosis (CVST), highlighting the role of magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) in patients with presumed idiopathic intracranial hypertension (IIH). Study was conducted in the department of neuro-ophthalmology at a tertiary eye care center in South India. Data from 331 patients diagnosed with IIH from June 2005 to September 2007 was included. Inclusion criteria were: Elevated opening cerebrospinal fluid (CSF) pressure of more than 200 mm of water on lumbar puncture, normal CSF biochemistry and microbiology, and normal neuroimaging as depicted by computed tomography (CT) scan. Exclusion criteria were: Space-occupying lesions, hydrocephalus, meningitis, intracranial hypertension (IIH). Study was conducted in the department of neuro-ophthalmology at a tertiary eye care center in South India. Data from 331 patients diagnosed with IIH from June 2005 to September 2007 was included.

Results

The clinical spectrum of CVST closely mimics that of idiopathic intracranial hypertension (IIH). IIH is mainly a diagnosis of exclusion based on Dandy’s criteria.1 Presence of elevated intracranial pressure (ICP) on lumbar puncture, normal cerebral spinal fluid (CSF) biochemistry and microbiology, no localizing sign with the exception of abducens nerve palsy and normal neuroimaging as depicted by computed tomography (CT) scan. Since Dandy’s criteria were formulated prior to the MRI era,2 CT scan alone may be an insufficient tool for accurate diagnosis of IIH. MRI, along with MRV wherever necessary should be the modality of neuroimaging for accurate diagnosis.3,4

Risk factors for cerebral venous sinus thrombosis (CVST) may not be apparent in all the cases, hence it is difficult to exclude CVST clinically in these patients. Magnetic resonance imaging (MRI) in combination with magnetic resonance venography (MRV) is recommended to correctly diagnose CVST in these patients.2,4

We report the rate of occurrence of CVST in patients with presumed IIH as well as the associated risk factor profile, which prompted subsequent MRV.

Materials and Methods

The study was conducted in the department of neuro-ophthalmology at a tertiary eye care center in South India. Patients diagnosed with IIH between June 2005 and September 2007 were included in this study. Inclusion criteria were: Elevated opening CSF pressure of more than 200 mm of water on lumbar puncture, normal CSF biochemistry and microbiology, and normal neuroimaging as depicted by CT scan.

Patients were excluded if they had space-occupying lesions, hydrocephalus, meningitis, ICP within normal range, abnormal CSF biochemistry and microbiology or bilateral disc edema from other causes like ischemic optic neuropathy, diabetic papillopathy. The remaining patients were evaluated with MRI and MRV. CVST was present in 11.4% of patients who were presumed to have IIH (35/308). MRI alone identified 24 cases (68%) of CVST, while MRI used in combination with MRV revealed an additional 11 cases (32%). Risk factors associated with CVST were identified in nine out of 35 patients (26%). CVST may be misdiagnosed as IIH if prompt neuroimaging by MRI and MRV is not undertaken. Risk factors of CVST may not be apparent in all the cases and these patients are liable to be missed if CT scan alone is used for neuroimaging, hence MRI, combined with MRV should be undertaken to rule out CVST.

Key words: Cerebral venous sinus thrombosis, idiopathic intracranial hypertension, magnetic resonance imaging, magnetic resonance venography

Indian J Ophthalmol: 2010;58:153-155

DOI: 10.4103/0301-4738.60092

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Patients were excluded if they had space-occupying lesions, hydrocephalus, meningitis, ICP within normal range, abnormal CSF biochemistry and microbiology or bilateral disc edema from other causes like ischemic optic neuropathy, diabetic papillopathy. The remaining patients were evaluated with MRI, and MRV. MRV was performed only when MRI was not undertaken. Risk factors of CVST may not be apparent in all the cases and these patients are liable to be missed if CT scan alone is used for neuroimaging, hence MRI, combined with MRV should be undertaken to rule out CVST.

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Discussion

The clinical presentation of CVST closely overlaps that of IIH. IIH is a diagnosis of exclusion according to Dandy’s criteria\(^1\) i.e. symptoms and signs attributable to the raised ICP, abnormal CSF pressure on lumbar puncture, normal CSF composition, normal neuroimaging results, awake and alert patient and absence of localizing findings on neurological examination.

The dynamics of ICP are described by the relationship:\(^5,6\)

\[
\text{Intracranial pressure} = \frac{\text{Cerebrospinal fluid formation}}{\text{Cerebrospinal fluid outflow resistance}} + \text{PSS},
\]

where

- Cerebrospinal fluid formation
- Cerebrospinal fluid outflow resistance, and
- PSS = outflow pressure in the superior sagittal sinus.

It is apparent from this relationship that sagittal sinus hypertension will cause a concomitant increase in ICP.

The type of neuroimaging has to be modified in view of associated risk factors for CVST, namely male sex, non-obesity, use of oral contraceptives, deep vein thrombosis, hypercoagulable states, lupus anticoagulant, infections of ear, nose and throat, mastoiditis and history of surgery in the region of head and neck. It is crucial to differentiate CVST from IIH because the management protocols are entirely different. CVST is typically treated with anticoagulants and IIH with diuretics. CVST is life-threatening as it can cause stroke and death, unlike IIH, which stops with optic atrophy.

The combination of MRI and MRV is the diagnostic modality of choice for CVST.\(^7,8\) MRV alone may be false-positive 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.\(^9\)

The dural sinuses most frequently affected are superior sagittal sinuses, transverse and sigmoid sinuses.

Our results differ from the study of Lee et al.\(^{10}\) a smaller study that reviewed MRV results in 22 consecutive patients, all women who fit the typical demographic profile of IIH, and identified none of the patients with CVST. Purvin et al.\(^{11}\) suggested that clinical manifestations of CVST may be differentiated from IIH by the abrupt onset and marked severity of symptoms.

However, the presentation of CVST can vary considerably based on the extent and location of thrombosis. The presentation of most of our patients with CVST was not the same as that described by Purvin et al. and closely mimics that of IIH.

Our study identified CVST in 11.4% of patients who were presumed to have IIH, and in the absence of correct diagnosis and lack of MRI and MRV these patients might continue to get treated as IIH, resulting in dismal visual prognosis, as well as significant risk of stroke and death. MRV in combination with MRI is recommended to identify this subgroup of patients who present with associated risk factors.

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