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Original Research Article

Cerebral sinus venous thrombosis

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ABSTRACT

Background: Intracranial Sino venous occlusive disease is an infrequent condition and accounts only 0.5-1% of all strokes. Objective of the study was to know the prevalence, pattern and risk factors involved in patients of cerebral sinus venous thrombosis (CSVT).
Methods: Hospital databases were searched retrospectively and patients diagnosed with CSVT from December 2014 to February 2016 were identified. Data on clinical presentation, risk factor, cerebral sinus involved, hospital stay and outcome were analyzed.
Results: Fifteen patients (3.4%) were identified as CSVT out of a total acute stroke case of 430. Mean age of patients was 31 years and 11 (73.3%) were males. The presenting symptoms included headache (n=14), altered sensorium (n=1) and seizures (n=5); signs included hemi paresis (n=4), papilledema (n=8) and bilateral VI nerve palsy (n=3). Majority had aseptic CSVT (n=14), and one patient had septic CSVT associated with tuberculous meningitis (TBM). Radiological findings included brain parenchyma involvement (n=8), subdural hematoma (SDH) (n=1), subarachnoid haemorrhage (SAH) (n=1) and normal brain parenchyma (n=5). Multiple cerebral sinuses were involved in seven patients. Most common site of CSVT was observed in sigmoid sinus. Risk factors included hyperhomocystenemia (n=3), protein S deficiency (n=8), protein C deficiency (n=6) and antithrombin III deficiency (n=3) as primary hypercoagulable state. ANA positive was observed in one patient and four had history of alcohol use. All fifteen patients received low molecular weight heparin (LMWH) for 1 week along with bridging oral anticoagulant. None of the patients required intrasinus thrombolysis and decompression surgery. Average hospital stay was 9 days. All had modified Rankin scale (MRS) ≤ 2 at 6 months follow up.
Conclusions: Prevalence of CSVT is higher than that reported from Asian studies. Younger age and male predominance was observed. Protein S deficiency was the major risk factor for CSVT.

Keywords: Cerebral sinus venous thrombosis, India

INTRODUCTION

Ribes et al first discovered CSVT in 1825 on autopsy cases. CSVT affects predominantly younger people with female gender predominance.[1] CSVT is considered to be a multifactorial condition associated with inherited and acquired risk factors. Inherited risk factors include hyperhomocystenemia, factor V Leiden mutation, protein C, protein S and antithrombin III deficiency and positive anti-cardiolipin and antiphospholipid antibodies.[1,2]

Acquired risk factors include central nervous system infections, head trauma, brain tumor, systemic vasculitis, hematological conditions, nephrotic syndrome, pregnancy and puerperium and dehydration. The purpose of present study is to investigate the prevalence, pattern and risk

factors involved in patients diagnosed with CSVT in present hospital.

METHODS

Hospital databases were searched retrospectively and patients evaluated thoroughly and diagnosed as CSVT at our Institute from December 2014 to February 2016 were identified.

Data on predisposing conditions, risk factors, clinical presentation, imaging findings, sinuses involved, duration of hospital treatment and outcome were collected.

All patients underwent detailed evaluation, that included, Computed tomography brain (CT), Magnetic resonance imaging (MRI) brain with venography, routine blood investigation (complete blood picture, erythrocyte sedimentation rate (ESR), blood sugar, renal function tests, liver function tests, antineutrophilic antibody (ANA), antiphospholipid antibody (APLA), Lupus anticoagulant (LA) and primary hypercoagubulable factors evaluation.

Protein C, protein S, antithrombin III, factor V Leiden, and homocysteine were routinely done in all the cases diagnosed as CSVT prior to anticoagulation therapy. Clinical outcome included mortality during hospital stay and functional outcome was analyzed by modified Rankin Scale (mRS) assessed at the time of admission, discharge and follow up at 6 months. Data was analyzed using SPSS version 17.

RESULTS

Fifteen patients were identified during the study period. CSVT accounted 3.4% of stroke. Age range of study group was between 18-47 years. Mean age of patients was 31.3±9.45 years of which 11 (73.3%) were males. Average hospital stay was 9.3 days.

Observation from present study is summarized in Table 1 (clinical presentation), Table 2 (risk factors), Table 3 (Imaging findings) and Table 4 (Dural sinus involved).

Table 1: Clinical presentation.

Symptoms	Number of patients (%)
Headache	14 (93.3)
Seizures	5 (33.3)
Vomiting	4 (26.6)
Altered sensorium	1 (6.6)
Hemi paresis	5 (33.3)
Signs	
Papilledema	5 (33.3)
Hemi paresis with seventh cranial nerve palsy	5 (33.3)
Sixth nerve palsy	3 (20)

Table 2: Risk factors and primary hypercoagulable state.

Risk factors	Number of patients (%)
Homocysteinemia	3 (20)
Diabetes mellitus	1 (6.6)
Alcoholism	5 (33.3)
Infection	1 (6.6)
Protein C deficiency	5 (33.3)
Protein S deficiency	6 (40)
Antithrombin III deficiency	3 (20)
Factor V Leiden mutation	Nil
ANA positive	1 (6.6)
No risk factors identified	3 (20)

Table 3: Imaging findings in CSVT.

Brain findings in MRI Brain	Number of patients (%)
Brain parenchyma involvement	8 (53.3)
Subdural hematoma	1 (6.6)
Subarachnoid hemorrhage	1 (6.6)
Normal brain parenchyma	5 (33.3)

Table 4: Dural sinus involvement in patients with CSVT.

Sinus involved	Number of patients (%)
Superior sagittal sinus alone (SSS)	3 (20)
Sigmoid sinus (SS)alone	5 (33.3)
SSS+bilateral transverse sinus (TS)	2 (13.3)
SSS+unilateral TS+ Unilateral SS	2 (13.3)
Unilateral SS+ Unilateral TS	3 (20)

Illustrative case 1

54-year male without prior medical illness presented with headache and right-sided hemi paresis.

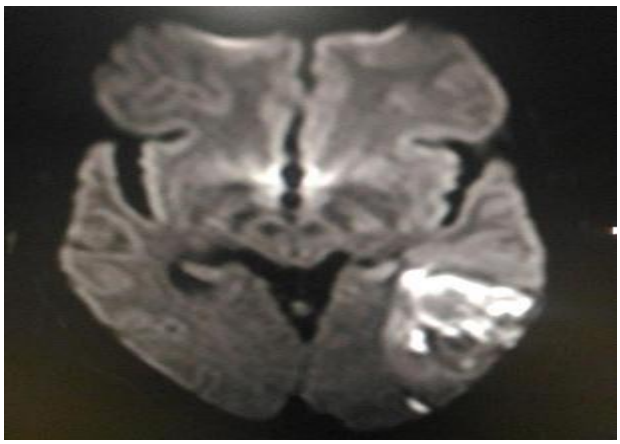

Figure 1: MRI brain diffusion images showing haemorrhagic infarct in left parieto-occipital region suggestive of venous infarct.

Magnetic resonance Imaging (MRI) brain showed diffuse restriction with hypo intensities suggestive of left parieto-occipital hemorrhagic infarct (Figure 1) with venogram brain showing left sigmoid and transverse thrombosis (Figure 2 and 3). Treated with LMWH followed by oral anticoagulant. Risk factor for CSVT was protein S deficiency.

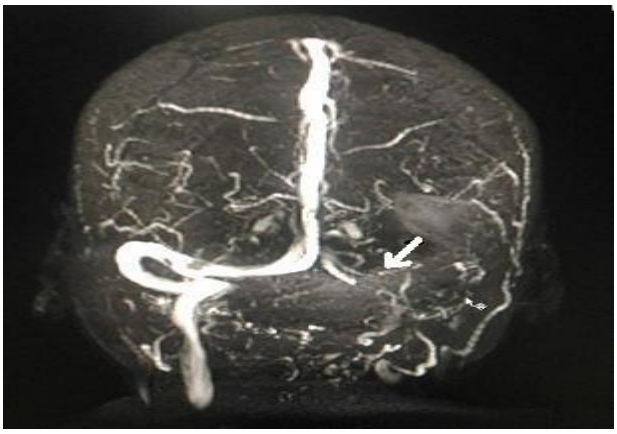

Figure 2: Thrombosis of left transverse sinus on venogram of brain.

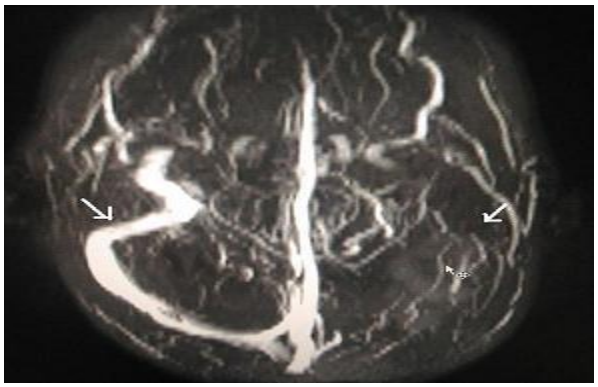

Figure 3: Left sigmoid sinus thrombosis marked by arrow on left side and right side normal sigmoid sinus.

Case 2

19 year female presented with acute onset of headache with vomiting of 1 week duration. MRI brain showed normal brain parenchyma with features of superior sagital sinus thrombosis evident on venogram brain (Figure 4). Risk factor was protein c deficiency without any predisposing illness of fever, dehydration and hormonal drugs. Recovered completely with anticoagulants and antiedema measures. Venogram brain MRI showing non visualization of superior saggital sinus (SSS) as marked by arrow in top row, subsequent images of brain with contrast showed empty delta sign marked by arrow in lowest row, suggesting SSS thrombosis. All the fifteen patients received LMWH for 1 week with bridging oral anticoagulant. Majority had aseptic CSVT (n=14) and one patient had septic CSVT secondary to TBM, who received antituberculous treatment with steroids in addition to anticoagulation for a short period. One patient developed carotid cavernous dural arteriovenous fistula at 3 months, presenting as proptosis of right eye with prominent conjunctival blood vessel that had protein S deficiency. He required endovascular intervention using onyx injection through venous route. MRS at 6 months follow up was ≤2 in all fifteen cases.

Figure 4: Venogram brain MRI showing non visualization of superior saggital sinus (SSS) as marked by arrow in top row, subsequent images of brain with contrast showed empty delta sign marked by arrow in lowest row, suggesting SSS thrombosis.

DISCUSSION

CSVT is more common in developing countries, usually affecting young individuals. CSVT accounts 0.5-1 % of all strokes.[1] In the present study frequency of CSVT was higher (3.4%) than that reported. Data from a monocentric cohort study over 15 years reported age range of 17-74 years with median age as 43 years.[2] In the present study purely younger population (18-47 years) were affected with CSVT with median age of 31 years lower than that reported. Most studies reported female sex preference in patients affected with CSVT, with no gender difference in children and elderly based on gender specific risk factors.[4-5] Kalita et al, Koopman et al and stated that CVST was more frequently associated with oral contraceptive use, pregnancy, or puerperium.[6,7] In present study male gender (73.3%) was predominantly affected with CSVT than females. This is attributed to the different risk factor involved in predisposing CSVT in the study group. The most common cause observed was primary hypercoagulable state. Secondly alcohol habituation (33%) was observed in male gender. Protein S deficiency was the most common disorder, followed by

protein C with Protein S and Protein C deficiency. Similar findings were reported by Chen WH et al.[8]

Haghighi et al studied 465 patients in the age group 29.5-43.8 years with ratio of female to male being 2.79.[9] Headache (80%-97%), sensory/motor deficits (39%-64%), and seizure (20%-62%) were the most common clinical presentations. Similarly our study had headache (93.3%) as the most frequent symptom in patients with CVT with 73.3 % being male population. The frequency of headache is higher than that reported by Mehndiratta et al, where headache (80%) was the most frequent symptom in patients with CVT with 75% being female gender.[10] Headache followed by seizures (33.3%) were the most frequent symptom observed in the present study. Similar findings have been reported by Nagarajan et al. Sigmoid sinus (SS) was the most commonly affected sinus either alone or in combination (66.6%) in present study. This differ from the prior studies reported by Nagarajan et al and Azin et al, were Superior sagittal sinus (SSS) was predominantly involved, 46% and 80.3% respectively.[11,12]

Literature review showed SSS thrombosis was the most common site associated with dehydration, sepsis, pregnancy, and puerperium. In the present study most common cause of CSVT was primary hypercoagulable state hence deeper sinuses were affected more than SSS. It emphasizes the evaluation of thrombophilic conditions to prevent recurrence of thromobosis. 53.3% of present study sample had involvement of brain parenchyma in form of infarct or hemorrhagic infarct, while 33.3% cases had normal brain parenchyma on MRI brain. A study from a French group identified 17 patients (13.8%) out of 123 in whom headache was the only manifestation of CSVT in the absence of focal deficit, or any parenchymal lesion.[13] Headache as sole manifestation with normal MRI brain was higher in present study (33.3%). This highlights evaluation of venous system of brain in patients presenting with isolated headache.

CONCLUSION

Present study provided details on the clinical manifestations, risk factors, neuroradiological findings, and outcome, of the patients with CVST in our hospital. Younger age of onset, male predominance, headache as the frequent symptom, with protein S deficiency being the commonest risk factor and major involvement of sigmoid sinus either isolated or combined, was observed in present study. Study highlights evaluation of primary hypercoagulable state in suspected cases of CSVT for decision regarding duration of anticoagulant therapy and to prevent future recurrence.

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Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

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