Case Report

Cerebral Venous Sinus Thrombosis: A Case Report

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Abstract

Cerebral venous sinus thrombosis (CVST) is a rare but dangerous condition occurring with an incidence of 3-4 cases per million per year. CVST presents a diagnostic challenge due to its varied presentation pattern. Here we report a case of a 40 year old diabetic, male, presenting with sudden severe headache with quadriplegia, where two predisposing factors were found.

Keywards: *Cerebral venous sinus thrombosis, Headache, ICU.*

Introduction

CVST is a rare category of stroke that can be promoted by a variety of conditions.[1-3] The most frequent symptoms and signs are headache (95%), focal seizure with or without secondary generalization (47%), unilateral and bilateral paresis (43%), and papilledema (41%).[4] The clinical presentation of CVST is varied and this creates a diagnostic challenge for clinicians.

Case report

A 40-year old diabetic male, was admitted in Intensive Care Unit (ICU) with the complaints of sudden onset of severe generalized headache for 2 days, followed by several episodes of vomiting, and weakness of all limbs for 1 day. He had difficulty in speech, but could communicate otherwise. There was no history of fever or trauma to the head. On examination, he was dehydrated, hemodynamically stable, conscious, had motor aphasia, both pupils were normal in size and reactive, no papilledema, upper motor neuron type of weakness of all four limbs with bilateral extensor plantar reflex. He had no signs of meningeal irritation or cerebellar lesion. After admission, he developed fever and hypotension. He was treated with adequate fluids, pressors and antibiotics. Patient gradually developed altered level of consciousness and had two episodes of convulsions, which were controlled by anticonvalsant. Laboratory investigations revealed high Hb% (16.2 gm/dl), HCT-52.6%, ESR was 50 mm in the 1st hour with neutrophilic leucocytosis. Random blood sugar-15 mmol/l, repeated urine dipstick examination showed persistent ketonuria. CT scan of Head showed diffuse cerebral edema.

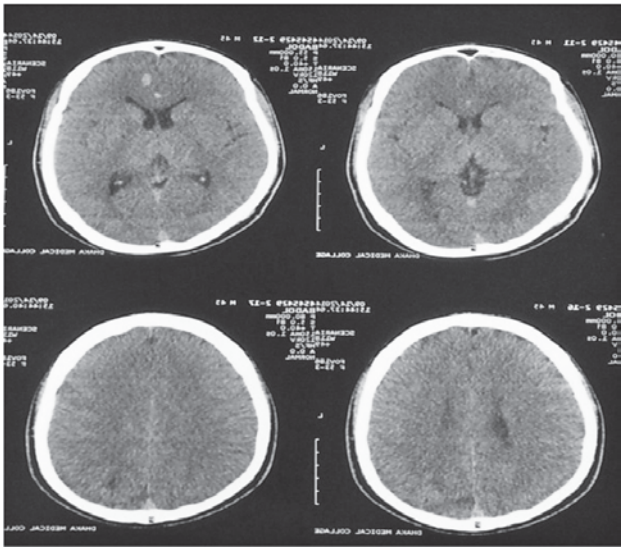

Figure 1: CT scan of head shows diffuse cerebral edema.

MRI brain showed diffuse altered signal areas in both hemispheres involving fronto-parietal regions with cerebral edema.

CSF study revealed total protein-63 mg/dl, glucose-12 mg/dl, total count-20 cells/cm^3, with neutrophil predominates (98%). ICT for malaria and febrile antigen test were negative, blood C/S, urine C/S showed no growth. Chest X-ray was unremarkable, Echo showed no regional wall motion abnormality, no vegetation, with good LV systolic function (EF-73%). D dimer was positive. Cerebral venous sinus thrombosis was suspected on MRI brain, Magnetic Resonance Venography (MRV) of brain was done which showed occlusion in posterior segment of superior sagittal

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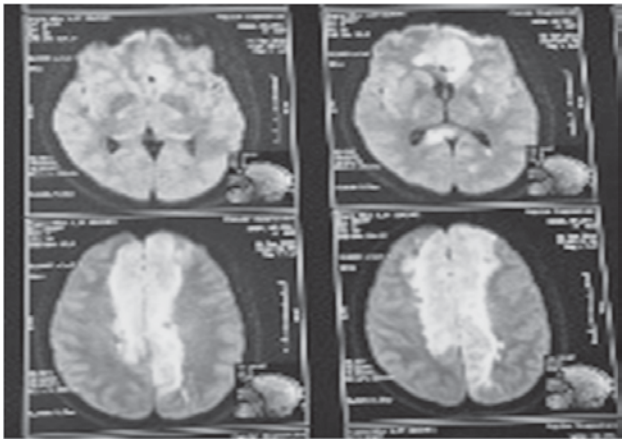

Figure 2: MRI Brain showed diffuse altered signal areas in both hemispheres involving fronto-parietal regions with cerebral edema.

sinus with hypoplastic left transverse sinus. Diagnosis of cerebral venous sinus thrombosis was established.

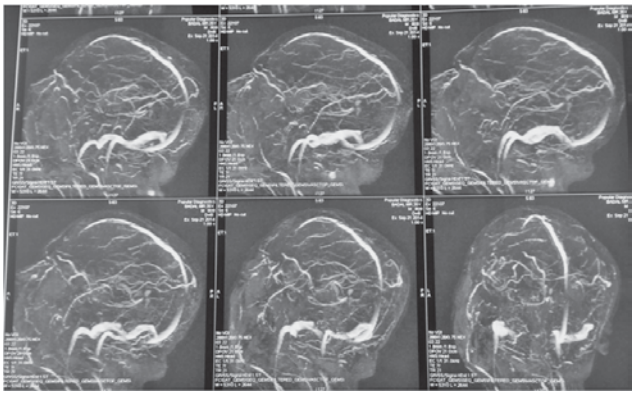

Figure 3: MRV of brain showed posterior segment of superior sagittal sinus.

Low molecular weight heparin was started. Vasculitis and coagulation abnormalities were evaluated. TPHA, VDRL, antibody for HIV 1, 2 were negative. ANA, p-ANCA, c-ANCA were also negative. Lipid profile showed total cholesterol 213mg/dl; LDL, HDL, triglyceride were normal. Serum homocysteine level was normal. Unfortunately due to financial constraints, patient was discharged against medical advice and ultimately died.

Discussion

The incidence of CVST is about 3-4 cases per million per year.[5] Women are affected three times more than men. Although majority of the patients gradually recover from acute episodes of CVST, one in eight patients suffers from either chronic disability or death.

Predisposing risk factors are found in 80%.[4] These may include prothrombotic condition,[4] head injury,[7] inflammatory diseases,[8] dehydration[9] and malignancy.[6,9] Our patient had dehydration and meningitis which was evident by positive CSF findings. His thrombophilic state evaluation so far done was normal.

According to the International Study on Cerebral Vein and Dural sinus Thrombosis, the most commonly affected site is the transverse sinus followed by superior sagittal sinus.[8] Other less common sites are the cortical vein, jugular vein and internal cerebral vein. In most patients, thrombosis occur in more than one sinus.

The variation in which CVST presents makes it a diagnostic challenge. Most frequent symptom is headache, tends to develop insidiously over days to weeks.[10] However, sudden onset with a thunderclap headache has been reported[11] which was noted in our patient. Rarely, a unilateral focal neurological lesion such as hemiparesis may develop, however this typically becomes bilateral in the following days. Seizures occur in 40% of patients and acts as a helpful differentiator between venous and arterial thromboses.[12] Our patient had quadriplegia and seizures. Raised intracranial pressure can lead to visual disturbances and papilledema on fundoscopy.[13]

Investigations are used in this condition in confirming the diagnosis and establishing any predisposing causes. Initially conventional CT scanning is usually performed to exclude other possible diagnoses such as subarachnoid haemorrhage. In 35% of published cases, CT scanning with contrast enhancement may demonstrate "empty delta sign" which is a useful radiological sign for the diagnosis of superior sagittal sinus thrombosis.[14] Furthermore, the use of transcranial color-coded duplex sonography is becoming increasingly common as a complement to other imaging techniques in the diagnosis of CVST.[15] The preferred and most sensitive diagnostic investigation is Magnetic Resonance Venography[16] which allows the venous occlusion to be identified along with any consequence such as cerebral edema and areas of venous infarction, as was present in our case. It has been suggested that CT venography detects CVST with equal accuracy and may be advantageous in allowing a prompt diagnosis immediately after initial CT scan[17]. When there is doubt following MRV, invasive cerebral angiography may be performed. Other investigations include lumber puncture and searching for thrombophilic state. CSF abnormalities noted in our patient are, increased protein content and neutrophilic pleocytosis suggestive of pyogenic infection. Thrombophilic state evaluation done so far in our patient was normal (protein C, protein S, antiphospholipid antibody could not be done due to financial constraints).

Management of patient with confirmed CVST include stabilization, anticoagulants, treatment and/or correction of the predisposing conditions. Despite the risk of haemorrhage into venous infarct, anticoagulation forms the mainstay of treatment, even in the presence of an existing haemorrhagic venous infarct[18], thus heparin is usually given after confirmation of the diagnosis. Our patient showed some clinical improvement following heparin administration. Warfarin is usually continued for 6 months, or longer in the presence of predisposing condition, with a target INR above 2.5 in order to reduce the risk of recurrent cerebral and extracranial thromboses.

The prognosis of CVST is usually favorable but is worse in extremes of ages, those with underlying conditions such as malignancy and sepsis, and in the presence of coma or deep cortical venous thrombosis.

Conclusion

CVST, due to the broad spectrum of clinical presentation, might be confused with other pathologies and so, frequently neglected. Thus, a high degree of suspicion is necessary for early recognition and instigation of appropriate therapy, thereby reduce mortality and morbidity.

References

1. Bousser MG, Chiras J, Bories J, Castaigne P. Cerebral venous thrombosis: a review of 38 cases. Stroke. 1985;16:199-213.
2. Gates PC, Barnett HJM. Venous disease: cortical veins and sinuses. In: Barnett HJM, Mohr JP, Stein BM, Yatsu FM, eds. Stroke Pathology, Diagnosis and Management. New York, Edinburgh, London, Melbourne: Churchill Livingstone; 1986: 731-743.
3. Lefkowitz D. Cortical thrombophlebitis and sinovenous disease. In: Toole JF, ed. Handbook of Clinical Neurology, Vol 54. New York/Amsterdam: Elsevier Science Publishing Co Inc; 1989: 395-423.
4. de Bruijn SFTM, de Haan RJ, Stam J, for the Cerebral Venous Sinus Thrombosis Study Group: Clinical features and prognostic factors of cerebral venous sinus thrombosis in a prospective series of 59 patients.J Neurol Neurosurg Psychiatry 2001, 70:105-108.
5. de Bruijn, SF, Stam J, Koopman MM and others. Case-control study of risk of cerebral sinus thrombosis in oral contraceptive users and in carriers of hereditary prothrombotic conditions. The Cerebral Venous Sinus Thrombosis Study Group. BMJ 1998; 316: 589-92
6. Ferro JM, Conhao P, Stam J, Bousser MG, Barinagarrementeria F: Prognosis of cerebral vein and dural sinus thrombosis: results of the International Study on Cerebral Vein and Dural Sinus Thrombosis (ISCVT).
7. deVeber G, Andrew M: Cerebral sino-venous thrombosis in children.N Engl J Med 2001, 345:417-423
8. Allroggen H, Abbott RJ: Cerebral venous sinus thrombosis. Postgrad Med J 2000, 76:12-15
9. Stiefel D, Eich G, Sacher P: Posttraumatic dural sinus thrombosis in children. Eur J Pediatr Surg 2000, 10:41-44
10. Ameri A, Bousser MG: Cerebral venous thrombosis. Neurol Clin 1992, 10:87-111
11. de Brujin, Stam J, Kapelle LJ: Thunderclap headache as a first symptom of cerebral venous thrombosis. CVST Study Group
12. Stam J: Thrombosis of the Cerebral Veins and Sinuses.N Engl J Med 2005, 352:1791-1798
13. Villringer A, Mehraen S, Einhaupl KM: Pathophysiological aspects of cerebral sinus venous thrombosis. J Neuroradiol 1994, 21:72-80
14. Lee EJ: The Empty Delta Sign.Radiology 2002, 224:788-789
15. Stolz EP: Role of ultrasound in diagnosis and management of cerebral vein and sinus thrombosis.Front Neurol Neurosci 2008, 23:112-121
16. Lafitte F, Boukobza M, Guichard J, Hoeffel C, Reizine D, Ille O, Woimant F, Merland J: MRI and MRA for diagnosis and follow-up of cerebral venous thrombosis (CVT).Clin Radiol 1997, 52:672-679
17. Smith R, Hourihan MD: Investigating suspected cerebral venous thrombosis. BMJ 2007, 334:794-795
18. Villringer A, Einhaupl KM: Dural sinus and cerebral venous thrombosis. New Horizons 1997, 5:332-341