Cerebral Venous Sinus Thrombosis : A Great Masquerader

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Abstract

Cerebral venous sinus thrombosis (CVST) is a rare form of stroke that results from thrombosis of the dural venous sinuses. While it may occur in all age groups, it is most common in the third decade with an estimated 3-4 cases per million annual incidence in adults. The disorder presents with varying clinical manifestations which may be nonspecific leading to a delay in diagnosis. If the patient presents with subarachnoid haemorrhage, the diagnosis may be more elusive. We present here a case of cerebral venous sinus thrombosis presenting as subarachnoid haemorrhage. 45 yrs male was admitted to our ward with history of 3 episodes of generalized tonic clonic convulsions. 2 episodes of vomiting and headache since 3 days. The convulsions were associated with frothing at the mouth and post ictal drowsiness. There was no history of weakness in any of the limbs. Evaluation included brain MRI which revealed a small sub-arachnoid haemorrhage (SAH) in the inter hemispheric fissure and absences of flow void in the superior sagittal sinus. An MR angiographic study of the brain revealed thrombosis of the superior/or sagittal sinus, left transverse sinus and left sigmoid sinus with sub-arachnoid haemorrhage (SAH) without any evidence of intra parenchymal bleed. After intravenous administration of unfractionated Heparin 5000 IU 12 hourly patient improved symptomatically. Follow up of MI scan after 7 days of IV heparin showed a partial resolution of the thrombus and no increase in size of the bleed. Warfarin was continued for 6 weeks.

Generally, in medicine, it has been shown that CVST is not treated with heparin. Present study observed an excellent results following European Federation of Neurological Societies guidelines.

Introduction

Cerebral venous sinus thrombosis (CVST) is a rare form of stroke that results from thrombosis (a blood clot) of the dural venous sinuses, which drain blood from the brain.[1]

Cerebral venous thrombosis (CVT) is an under diagnosed condition for acute or slowly progressive neurological deficit. CVT is less frequent than arterial thrombosis. CVT has a wide spectrum of signs and symptoms, which may evolve suddenly or over the weeks. It is clinically

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challenging and mimics many neurological conditions such as, meningitis, encephalopathy, benign intracranial hypertension, and stroke. With increasing awareness, CVT cases are now being diagnosed more frequently. Newer imaging procedures have led to easier recognition of venous sinus thrombosis, offering the opportunity for early therapeutic measures. It may be difficult to diagnose it on clinical grounds alone.[2]

While it may occur in all age groups, it is most common in the third decade with an estimated 3-4 cases per million annual incidence in adults.[3]

Given that older studies show no difference in incidence between men and women, it has been suggested that the use of oral contraceptives in women is behind the disparity between the sexes.[1] CVST is increasingly detected in modern times with the advent of greater awareness amongst clinicians and availability of noninvasive diagnostic modalities like magnetic resonance imaging and magnetic resonance venogram.[1] The disorder presents with varying clinical manifestations which may be nonspecific leading to a delay in diagnosis. If the patient presents with subarachnoid haemorrhage, the diagnosis may be more elusive.

We present here a case of cerebral venous sinus thrombosis presenting as subarachnoid haemorrhage. Till date 15 such cases are reported in literature.

Case Report

45 yrs male was admitted to our ward with history of 3 episodes of generalized tonic clonic convulsions. 2 episodes of vomiting and headache since 3 days. The convulsions were associated with frothing at the mouth and post ictal drowsiness. There was no history of weakness in any of the limbs. The patient is an alcoholic since last 20 yrs (1 bottle of country liquor /day). However he is a non smoker. He is non diabetic, non hypertensive, and had no evidence of any trauma and did not give any history of fever.

On examination; he was conscious, oriented in time, place and person. Pulse was 80/min, BP 130/80 mm Hg. There was no pallor, icterus or cyanosis. His higher mental functions were normal, power in all 4 limbs was grade 5, pupils were bilaterally normal in size and reacting to light, plantars were flexor. No cardiac respiratory, GI symptoms, no evidence of any source of infection of the ear, face or scalp.

On investigation : Hb was 11.5 gms%, total leucocyte count was 7600/mm^3 and platelet count was 150000. Liver and renal profile was normal. Fundoscopy did not reveal any papilloedema. Prothrombin was 15 sec, INR was 1.4, APTT was 29 secs. Homocysteine levels were raised. HIV and HBSAg were non reactive. Protein C, protein S and factor 5 laden were normal.

In view of these findings, a differential diagnosis of intracerebral bleed, stroke, cerebral malaria, meningitis was thought of. Further evaluation included brain MRI which revealed a small sub-arachnoid haemorrhage (SAH) in the inter hemispheric fissure and absences of flow void in the superior sagittal sinus. An MR angiographic study of the brain revealed thrombosis of the superior sagittal sinus (Fig. 1), left transverse sinus and left sigmoid sinus with sub-arachnoid haemorrhage (SAH) without any evidence of intra parenchymal bleed. At

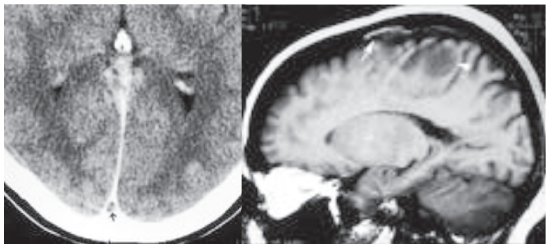

Fig. 1 MRI shows thrombosis of superior sagittal sinus

this point of time our major concern was to treat thrombosis with anticoagulants to decrease the raised intracranial tension without increasing the haemorraghe. Other medications included syrup glycerol and anticonvulsants. Intravenous unfractionated Heparin 5000 IU 12 hourly was administered. APTT as well as sensorium was monitored. The patient improved symptomatically. Follow up of MI scan after 7 days of IV heparin showed a partial resolution of the thrombus and no increase in the size of the bleed. Warfarin was continued for 6 weeks.

Discussion

We present here a case of cerebral

venous sinus thrombosis presenting as subarachnoid haemorrhage. The diagnosis was based on magnetic resonance imaging (MRI) employing radiocontrast to demonstrate obstruction of the venous sinuses by thrombus.[1]

Symptoms included tonic and clonic convulsions, headache and vomiting but absence of weakness in four limbs, abnormal vision, which is one of symptoms of stroke.

After 7 days of intravenous anticoagulation therapy, his condition stabilized. Similar finding has been reported by Lin et al in case of a 44-year-old man with superior sagittal and transverse sinus thrombi, who initially presented with venous subarachnoid haemorrhage over the right parietal sulci.

Repeated imaging after 7days revealed decrease in size of haemorrhage. Hence anticoagulants were commenced.[4]

We have followed guidelines of European Federation of Neurological Societies[3] regarding the use of anaticoagulants in case of CVST with SAH.viz.

1. Low miolecular weight heparin/unfractionated heparin is recommended face of SAH and is not contraindicated.
2. The use of thrombolytics either local or systemic lacks evidence based and is to be used only if anticoagulants fail.
3. Oral warfarin is to be used for 3 months for patients with thrombotic risk factor and for 6-12 months for those without a risk factor.
4. Anticonvulsants are to be used prophylactically.
5. Patients with vision failure should undergo a lumbar puncture to decrease the intracranial tension prior to anticoagulation.
6. Medical management of raised intracranial tension should include intubation, hyperventilation and osmotic diuresis.

Till date only two randomized control trials, one using unfractionated and the other with low molecular weight heparin have been documented. [5,6]

Chang *et al*[7] reported three cases of isolated cortical venous thrombosis (CVT) presented with unilateral, localized subarachnoid haemorrhage without parenchymal involvement. (CVT) without concomitant dural sinus thrombosis is an uncommon disorder. Isolated CVT usually manifests on imaging studies as focal parenchymal haemorrhage or oedema.

In the present case homocysteine levels were raised but lipids were in normal limits. However, out of 6 patients of cerebral venous thrombosis (490 stroke registry) documented. hyperhomocysteinaemia and hyperlipidaemia in one male by Mehndiratta *et al* from Delhi.[2]

Mehndiratta *et al* from Delhi[2] documented 6 patients of cerebral venous thrombosis out of 490 stroke registry. Of these 6, four were females and two were males. The mean age among females was 27.75 years and among males was 41.5 years. Of the 4 females two were postpartum; one was on oral contraceptive and one on Antiphospholipid antibodies (APLA) positive. Amongst two males one had hyperhomocysteinaemia and one had hyperlipidaemia.

Heparin 5000 IU 12 hourly was administered in the present study and treatment was continued for six months since underlying cause of condition was unprovoked but there are no clear causes or a "mild" form of thrombophilia, 6 to 12 months is advised.[3]

Clinical practice guidelines now recommend heparin or low molecular weight heparin in the initial treatment, followed by warfarin, provided there are no other bleeding risks that would make these treatments unsuitable.[8, 9, 10]

In view of generalized tonic and clonic convulsions anticonvulsants were used to prevent seizures.[3]

Nondiabetic and nonhypertensive status of our subject was an unusual presentation though CVST in a 26 year old female with diabetic ketoacidosis has been documentated by Mohamad. *et al.*[11]

Dural venous sinus thrombosis may present to the physician in a number of guises. Diagnosis can be confirmed by MR imaging in most cases. Early recognition of the condition and instigation of appropriate therapy probably reduces mortality and morbidity. The initial treatment should be intravenous heparin, with thrombolysis reserved for cases undergoing secondary deterioration.[12]

Cerebral venous thrombosis can be difficult to diagnose and is further complicated when patients initially present with acute subarachnoid haemorrhage. A high index of clinical suspicion is needed to diagnose venous subarachnoid haemorrhage so that appropriate treatment can be initiated as promptly as possible,[13] resulting in excellent long-term prognosis with full recovery, can be achieved.[14]

Generally, in medicine, it has been shown that CVST is not treated with heparin. Present study observed an excellent results following European Federation of Neurological Societies guidelines.

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