Cerebral Venous Sinus Thrombosis : A Great Masquerader

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Introduction

Cerebral venous sinus thrombosis (CVST) and cortical venous thrombosis (CVT) leading to ischemia and infarction of the brain are being increasingly detected in recent times with the advent of greater awareness amongst the clinicians and greater availability of non-invasive radiodiagnostic modalities like magnetic resonance imaging (MRI) and MR venography. Patients often present with protean neurological manifestations in the form of focal neurological deficits, seizures, headache and other features of raised intracranial tension, leading to misdiagnosis and delay in treatment. The clinical presentation may be with features suggestive of transient ischemic attack (TIA) or at times an intracranial space-occupying lesion (IC SOL). High index of suspicion is required in appropriate clinical settings to diagnose this entity at the earliest and ensure prompt treatment with anticoagulants, which can result in almost complete neurological recovery. We report three such cases, which presented to us with varying manifestations, and briefly discuss this entity.

Case Report-1

A 44 year old chronic alcohol abuser and smoker, presented with complaints of acute onset weakness of the right lower limb lasting for a day followed by weakness progressively involving the right upper limb and the face over the next 24 hours with partial recovery of the weakness over the next 3 days. The illness was preceded by headache and 5-6 episodes of recurrent generalised tonic-clonic seizures (GTCS), each lasting 3-4 minutes. He denied any speech abnormality, cranial nerve deficit, sensory symptoms or sphincter incontinence. There was no history of fever, vomiting, blurred vision, ear discharge or head injury. He denied past history of any major illness or symptoms suggestive of TIA.

On examination, he was afebrile with pulse 88/min, regular, blood pressure of 140/80 mmHg and respiratory rate of 16/min. There was no pallor, icterus, tremors, diaphoresis, or

signs of hepatic dysfunction. Neurological evaluation revealed a confused male having grade III/V power in right upper and lower limbs with spasticity of both lower limbs and bilateral extensor plantars. There was no papilledema, sensory deficit, ataxia or signs of meningeal irritation. Examination of the other systems was normal.

Investigations revealed Hb 11.8g/dL, macrocytosis (MCV 104fl), normal total and differential leukocyte and platelet counts, normal liver and renal functions and blood sugar levels. Chest radiograph and ultrasound of the abdomen revealed no abnormality. Coagulation profile (prothrombin and activated partial thromboplastin time) was within normal limits. NCCT brain showed bilateral parietal lobe hemorrhagic infarcts. MRI of the brain showed evidence of superior sagittal sinus (SSS) thrombosis and venous infarcts in both parietal lobes with minimal central hemorrhagic transformation (Fig 1a - sagittal plane) and (Fig 1b - coronal plane). Work up for primary thrombophilic state was planned after three months.

The patient was managed with parenteral vitamins (vitamin B1, B6 & B12), subcutaneous low molecular weight (LMW) heparin (Enoxaparin) 40mg SC twice daily for seven days and tablet phenytoin sodium 300mg at bedtime. Oral anticoagulation (Tab Acinocoumarol 2mg OD) was started simultaneously and optimized at International Normalised Ratio (INR) of 2 following which heparin was withdrawn. His neurological deficit recovered gradually and there was no recurrence of seizures. He was discharged on day 15 of illness without residual neurological deficit and was advised to continue anticoagulation for three months.

Case Report-2

A 52 year old male presented with episodic occipital headache associated with vomiting and blurred vision of one-week duration. This was associated with transient weakness of left half of the body and later right half of the body, each lasting for a few hours, two days prior to hospitalisation. He denied having seizures or altered sensorium. He had one episode of similar headache one month prior to hospitalisation, which subsided spontaneously. On examination, he was found to have bilateral papilledema, both lower limb spasticity

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[1]. In about 25% of patients, no predisposing risk factor can be discovered despite extensive investigations. In the three cases which have been presented, no definite cause could be found on initial workup.

CVST most commonly involves SSS (72%) followed by lateral sinus (70%). In 30-40% cases, more than one sinus is involved with or without cortical venous thrombosis [2]. The sinuses involved in our cases were SSS in case no.1 and 3 and SSS and transverse sinus in case no. 2. Clinical manifestations seen in CVST are headache (75-90%), papilledema, focal neurological deficits (75%) and seizures [1]. Rarely, about 4% of patients with SSS thrombosis present with bilateral or alternating lower limb weakness as in case no. 2. Unusually, patients present with a "thunderclap headache" mimicking subarachnoid hemorrhage (SAH) [4].

Neuroimaging modalities of choice in CVST are computerized tomographic (CT) scan and MRI with MR venogram (MRV). CT scan may be normal in 15-30% cases but MRI with MRV is almost 100% diagnostic [5]. CT scan pictures suggestive of CVST are infarcts, which may not conform to an arterial territory, extensive cerebral edema, hemorrhagic infarcts, falcine and tentorial enhancement and thrombosed cortical vein (Cord sign). MRI with MRV is the investigation of choice, which shows absence of flow void in the thrombosed sinuses [5]. Newer modalities for diagnosis include the flow-sensitive imaging techniques like the 2D time-of-flight and the phase-contrast MRV which have a higher sensitivity in picking up the venous thrombi. In all our cases, there was conclusive evidence of CVST on MRI with MRV. Cerebrospinal fluid analysis is important to exclude important differential diagnosis of meningitis and SAH in the appropriate clinical settings [6].

Work-up for underlying primary thrombophilic state is ideally postponed to 8-12 weeks after the acute episode as these tests may be falsely negative or positive in the acute state, and the results are also affected by the administration of anticoagulant [7]. Anticoagulants must be withdrawn at least for 2 weeks prior to testing for these abnormalities. The cause for the recent surge in cases of CVST, especially with reference to our patients is yet to be ascertained though there is a temporal relation with hot summer days (temperature rising to above 40°C, noticed during the study period viz May-June 2002) and alcohol intake, which may have contributed to dehydration and a state of hyperviscosity.

Anticoagulation is the cornerstone of treatment in CVST. It is only withheld in patients with septic

thrombophlebitis and in large haemorrhagic venous infarct with significant mass effect. Oral anticoagulants should be overlapped with heparin and the latter discontinued once therapeutic INR of 2-3 is achieved. Oral anticoagulants should be continued for at least 3 to 6 months if no underlying procoagulant state is found, or life-long if there is an irreversible procoagulant state. In case no.1 & 2, anticoagulation was administered, as the haemorrhage was minimal and there was no mass effect but was withheld in case no.3 due to significant haemorrhage with midline shift. Thrombolysis of the clot using local administration or systemic injection of urokinase or recombinant tissue plasminogen activator has been evaluated and has been found to be effective though it is fraught with danger of hemorrhagic transformation of the infarcts and can be used only in centers equipped with facilities for intensive neurological monitoring [8].

CVST is a great masquerader as it can present in various forms and confuse the clinician. It manifests as a stroke with seizures confusing it with arterial strokes. It may present with headache and papilledema, the clinical sine qua non of ICSOL. It may also present with pleomorphic seizures occasionally with fever suggesting encephalitis. Hence, it requires an astute clinical sense tempered with diligent search to enable early diagnosis, ensure aggressive use of anticoagulants and lessen the morbidity and mortality in these cases.

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