Cerebral Venous Sinus Thrombosis: Our Experience with an Illustration at National Institute of Neurological and Allied Sciences

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Cerebral venous sinus thrombosis (CVST) is one of the relatively rarer conditions in neurosurgery. It presents with various neurological symptoms. It is often missed in usual neuroimagings and prognosis is often poor due to misdiagnosis and improper treatment.

Here we present a case of cerebral venous sinus thrombosis who presented with headache. Initially that was thought to be low pressure headache due to lumbar puncture 2 weeks back when he underwent appendicentomy under spinal anesthesia. Since his headache didn't improve further investigations were performed with enhanced computerized tomography (CT) and magnetic resonance venography (MRV). Finally CVST was diagnosed and treated accordingly.

Though CVST is relatively rare, it is being diagnosed and treated more frequently at National Institute of Neurological and Allied Sciences (NINAS), Bansbari, Kathmandu, Nepal.

Key words: CVST, Neuroimagings, NINAS

Cerebral Venous Sinus Thrombosis (CVST) is an uncommon medical condition which may present with a variety of symptoms ranging from headache, altered sensorium, opthalmoplegia, focal neurological deficits, hemiparesis, tingling sensation of limbs etc. Predisposing conditions implicated in this condition are low flow state of blood, inflammatory states of blood vessels, dehydration, fever, infection of the surrounding structures, pregnancy, use of oral contraceptive pills, hypercoaguble states etc. CSVT is not a common condition, its annual incidence in adults being 3-4 cases per million. It is common in women of third decades of age. Here we present a case of CSVT with atypical history and presentation.

Case report

A 25 year old gentleman presented to National Institute of Neurological and Allied Sciences, Bansbari Emergency room with the complaint of low grade intermittent generalised persistent headache for twelve days along with fever for four days. He developed altered sensorium for two days. He was admitted for appendicitis and underwent appendicectomy under spinal anesthesia fifteen days back at another hospital in Kathmandu. Subsequently he developed headache on the third post-operative day which was treated as postural hypotension with no relief. He is non smoker, non-alcoholic and had Pulmonary TB treated completely during childhood. There is no history of drug abuse, travelling abroad, trauma, use of anabolic steroids.

He was disoriented at presentation with eyes opening to speech and localising to pain. Pupils were 3mm in size bilaterally reactive to both direct and consenseual light reflex. Cranial nerves were intact. There were no focal neurological deficits. Fundus examination was normal. Neck rigidity was present but Kernig's sign was negative. Other systemic examinations were within normal limit.

He was admitted and all routine investigations were done. Comple blood count was 13,000/cumm with N73%L17% , hemoglobin was 12gm %, Bleeding time and

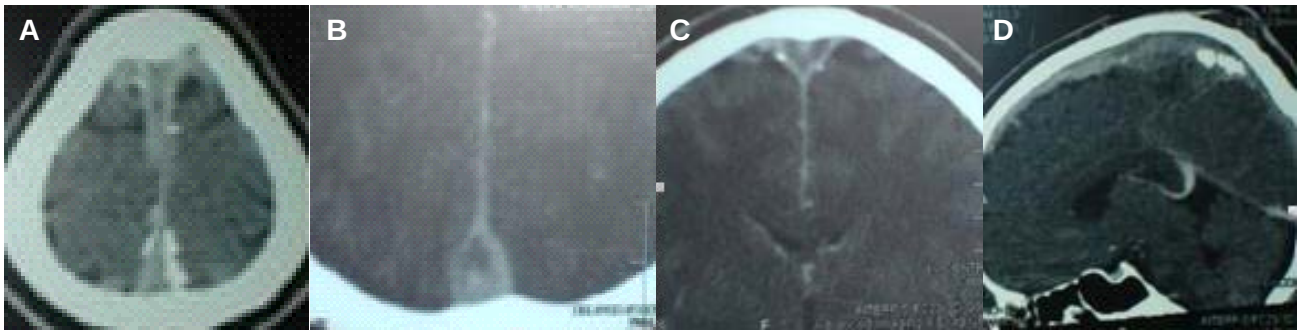

Figure 1. Computerized tomography scan with contrast enhancement showing thrombus in the occipital end of superior sagittal sinus (SSS), A) axial image, B) occipital end of SSS showing empty delta sign, C) coronal image and D) sagittal image

Clotting time was within normal limit. Platelets count was 2,30,000 cells/cumm Coagulation profile -PT 17sec, INR 1.1 at the time of admission. Electrolytes were within norml limit Chest X-ray was normal. CSF study was done which revealed pressure of 23cm of water, total count of 5 WBCs and RBC 8 with 85% neutrophils 15% lymphocytes, Sugar 54mg% protein 21 mg% Gram stain and AFB stain showed no organism and no growth on culture.Thease features of CSF study ruled out meningitis. D-dimer quantification was done which was 1.03 microgram /ml (normal 0.0 to 0.5 microgram/ml). Anticardiolipin antibody was within normal range. CT Scan of his brain was done which showed features of bilateral frontal sinusitis and low attenuating thrombus within superior saggital sinus surrounded by the triangular area of enhancement (Empty delta sign), there were no signs of hemorrhage, ischaemic changes, no space occupying lesion. Susbsequently MRI with MR Venogram was done which showed occlusion of superior saggital sinus.

So the diagnosis of Sinus venous thrombosis was made and treatment was started with initial heparinization for five days then he was subsequently managed with oral anticoagulant(Warfarin). Regular monitoring of PT/INR was done. Patient showed gradual recovery from the symptoms once the therapy started and completely recovered within one week while anticoagulation was continued for next six months. Dehydration induced by vomitting and fever, anorexia leading to poor oral intake of fluid due to appendicitis may have contributed along with lumbar punture with loss of CSF and injection of anesthetic agent into the brain circulation in the above case which responded quickly to fluid replacement and anticoagulation.

Discussion

Reliable data about the incidence of CSVT are not available but it is being diagnosed more in numbers at National Institute of Neurological and Allied Sciences, Bansbari during the last two years. As it present with highly variable clinical presentation, the diagnosis of cerebral venous sinus thrombosis (SVT), as a rare but important causes of stroke is challenging. Most of the cases presented with headache, altered sensorium, some neurological deficits like opthalmoplegia, limb weakness or tingling sesation recovered well with the convetional therapy with Heparinization and Warfarin. Review of the literatures from different studies showed mortality rate ranging between 20-80 % . Poor prognostic factors include late diagnosis, presence of papilloedema, coma or altered sensorium, elderly patient, intracerebral hemorrhage and involvement of Straight Sinus. Surviving patient may be left with residual paresis, seizure disorder, cognitive impairement, visual impairment, recurrence of CSVT or venous thrombosis elsewhere, incomplete re-canalization of venous sinuses, persistent raised ICP, venous infarction and hemorrhage, hypopituitarism, development of dural A-V fistulas, seizure disorder. Superior saggital and lateral sinuses are most commonly involved but all other cortical and cerebellar veins may be involved.

Literature reviews have shown more than hundred recorded causes of CSVT but most common were low flow states of blood, dehydration, hyperemesis, diarrhoea, infection of the brain surrounding structures including meningitis, middle ear infection, mastoiditis and facial skin, pregnancy, use of hormonal/ contraceptive pills, androgens, hypercoagulable states, hereditory pro-thrombotic states such as Factor V leiden, deficiency of Protein S,C and antithrombin III penetrating head trauma, nepthrotic syndrome etc.

CT Scan of the brain may be suggestive but MR Venogram of the brain is the most effective tool of diagnosis of suspected CSVT as it is difficult to diagnose the condition only from the clinical features. Because noncontrast cranial CT (NCCT) is still the imaging technique of choice in most of the neurological settings it should be focused on. Noninvasive Magnetic resonance (MRI/MRV) and computed tomography (NCECT/ CECT) and CT venography have largely replaced conventional angiography for initial evaluation and follow-up. These techniques have high sensitivity for diagnosing CVT. However, they also have pitfalls that can lead to false-positive and -negative results for example congenital hypoplastic veins may be misataken for CSVT in MRI. CT Scan Axial/saggital/coronal views of brain may demonstrate high attenuation presence of thrombus along the course

of Sagittal/ transverse sinuses. Filling defects within the course of venous sinuses in contrast enhanced CT (CECT) venogram are diagnostic of CSVT. Low attenuating thrombus within superior sagittal sinus surrounded by triangular area of enhancement (Empty delta sign) is diagnostic. Cerebral odema and venous infarction are also usual findings. D-dimer quantification is very useful investigation with high sensitivity and specificity for diagnosis of CVST and monitoring effectiveness of the anticoagulation.

Treatment of CVST is started with thrombolytic therapy with Heparin or low molecular weight heparin initially then maintaining on Warfarin. The duration of Warfarin therapy depends on the circumstances and cause of CVST ranging from 2 months to life-long therapy with regular PT/INR monitoring. Presence of anti cardiolipin antibody necessitates life-long anticoagulation therapy. Invasive thrombolysis by injecting thrombolytic agents like streptokinase is also done but reserved for the cases not responding to systemic anticoagulation therapy. Unwanted bleeding into brain or other structures may complicate the treatment. ICP lowering agents and anti-epileptic drugs are also added according to the need. Dietary modification is needed with restrictions of green vegetables, cabbages and liver.

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