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Cerebral venous sinus thrombosis: A diagnostic challenge in a rare presentation

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Abstract:

Cerebral venous sinus thrombosis (CVST) is an uncommon, life-threatening condition with a variable clinical presentation that makes it a challenge of diagnosis. A 39-year-old male patient presented to the hospital with complete loss of conscious and admitted to Medical Intensive Care Unit for investigation without any obvious history that was difficult for diagnosis. In this case, the patient presented with coma that is a rare presentation of CVST with no obvious clinical history and he was male patient that means he is free of all gender-specific risk factors of CVST. The brain computed tomography (CT) scan showed hypodense lesion in the left upper parietal region with no hemorrhage. The lesion was low-signal intensity (SI) on T1WIs and high SI on T2WIs and restricted on diffusion-weighted images like arterial infarctions, but magnetic resonance angiography (MRA) was normal that excluded arterial infarction. Gadolinium-enhanced MR venography (MRV) showed the filling defect of CVST. CVST can be present by a mysterious clinical presentation that makes it as a challenge of diagnosis even by medical imaging by CT and MR imaging (MRI). A combination of MRI and MRV is the best, noninvasive, and nonionizing imaging modality for the diagnosis of CVST.

Keywords:

Cerebral venous sinus, challenge, diagnostic, presentation, rare, thrombosis

Introduction

erebral venous sinus thrombosis (CVST) is the formation of a blood clot in the dural venous sinuses, which drain the blood and cerebrospinal fluid (CSF) from the brain to the internal jugular vein.[1]

CVST is an uncommon, life-threatening condition that needs early diagnosis and therapy. It has an extraordinarily variable clinical presentations range from headache to seizures and even coma that make it as a challenge of diagnosis and often not diagnosed clinically at presentation.[2]

It can affect any age group even neonates with a younger age at distribution than arterial stroke. It affects females more than

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males, and it commonly affects the superior sagittal sinus (SSS) and/or the transverse sinuses.[3]

Known risk factors of CVST may be either acquired such as oral contraceptive pills (OCPs), pregnancy, puerperium, and dehydration or genetic such as inherited thrombophilia.[4]

In the first 5 days, the venous thrombus appears as moderate signal intensity (SI) in T1WIs and low SI in T2WIs but from 5th to 15th day, the venous sinus thrombus appears as high SI in both T1 and T2WIs, and it can be diagnosed easily. MR venography (MRV) will show no flow in the veins since the 1st day of thrombosis.^[5]

Magnetic resonance imaging (MRI) with MRV is the imaging modality of choice to diagnose CVST.[6]

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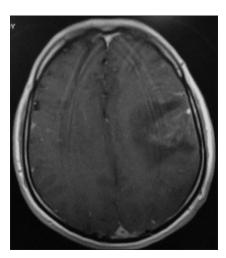


Figure 1: Axial gadolinium-enhanced T1 shows a filling defect in the superior sagittal sinus represents positive empty delta sign of superior sagittal sinus thrombosis with heterogeneous nonenhancing lesion in the left upper parietal region represents venous infarction

Anticoagulant drugs such as intravenous heparin are the main treatment of acute venous sinus thrombosis to prevent propagation of the thrombus, pulmonary embolism, and decrease the risk of death or dependency even if there is intracerebral hemorrhage (ICH).^[7]

The aim of this study is to document a case of CVST with a rare clinical presentation that was deceitful and led to do a false provisional diagnosis as an encephalitis and heavy clinical and medical imaging workup to reach the final diagnosis.

Case Report

A 39-year-old male patient presented to the hospital with complete loss of conscious that preceded by headache since the previous day. The patient admitted to the Medical Intensive Care Unit as a case of coma for investigation. Medical history and clinical examination gave no obvious cause of coma. Routine laboratory investigations including complete blood count and random blood sugar revealed unremarkable results. Brain computed tomography (CT) scan showed an ill-defined hypodense area in the left upper parietal lobe that suggested encephalitis or infarction as a diagnosis. MRI showed low SI area in the left upper parietal lobe with effacement of the cortical sulci and thickened gyri that were high SI on T2 and fluid-attenuated inversion recovery (FLAIR). These findings also suggested the diagnosis of encephalitis, but the patient was afebrile and not immunocompromised. Diffusion-weighted (DW) was requested to exclude cerebral infarction that showed diffusion restriction of the lesion and suggested infarction as a diagnosis. MR angiography (MRA) was normal and not compatible with an arterial infarction.

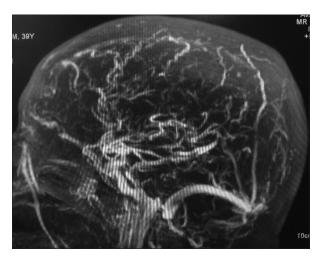


Figure 2: Sagittal magnetic resonance venography shows filling defect in the superior sagittal sinus represents superior sagittal sinus thrombosis

On the next day: Gadolinium-DTPA injection was given, T1WIs showed a filling defect in the SSS that represent the "cord sign" of sagittal sinus thrombosis [Figure 1]. Gadolinium-enhanced MRV showed a persistent filling defect in the SSS on the sagittal images [Figure 2] and also in the left transverse and sigmoid sinus on the axial images [Figure 3] that confirmed the diagnosis. The final diagnosis was SSS, left transverse and sigmoid sinus thrombosis with left parietal lobe venous infarction. Anticoagulant drugs were given to this patient and improvement achieved.

Discussion

CVST has different presentations that make it as a challenge for the clinical diagnosis. A headache is the predominant clinical feature (approximately in 92% of cases) as reported by Azin and Ashjazadeh^[1] In the current case, the patient presented with coma that is a rare presentation and commonly led to the false provisional diagnosis.

CVST is more common in females than in males with approximately 3:1 ratio and gender-specific risk factors such as OCPs, pregnancy, and puerperium are the most common important factors in the affected females.^[8,9] The current case was male patient and free of all these gender-specific risk factors.

The patient, in this case, had no obvious cause of CVST that is compatible with Saadatnia *et al.*^[10] who reported that approximately 30% of CVST has no underlying cause can be identified.

Pongmoragot and Saposnik^[11] reported that nearly third of cases of CVST present with ICH. In the patient of this study, brain CT-scan study excluded any cerebral hemorrhage.

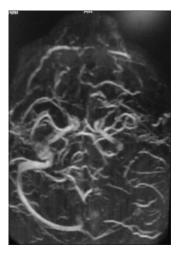


Figure 3: Is axial magnetic resonance venography shows filling defect in the left transverse and sigmoid sinuses represents venous thrombosis

Arterial infarction was a priority for diagnosis in the current case because of CVST is a rare cause of cerebral infarction in comparison of an arterial origin that is common as Choe *et al.*^[12] reported.

In this case, MRI showed low SI in T1WIs that was high SI in T2WIs and FLAIR with another small lesion in the right cerebral hemisphere that was restricted area on DW images. These findings were more consistent with a diagnosis of cerebral arterial infarction, but MRA was normal. MRV was done that showed a filling defect in the SSS in sagittal images and a filling defect in the left transverse and left sigmoid sinus in the axial MRV. The "cord sign" of SSS thrombosis was seen in axial images of Gadolinium-enhanced MRV. The above findings are compatible with Bousser and Ferro^[13] who reported that MRI alone is not enough to diagnose CVST because of the absence of high SI on T1 and T2 WIs at the first 3–5 days of thrombosis that appears of moderate SI on T1 and low SI on T2 WIs as the normal vein. They reported that MRV alone could not differentiate between thrombosis and hypoplasia of the transverse sinus and a combination of MRI sequences with MRV are the gold standard to diagnose CVST.

Findings in the current case confirm that MRV is the investigation of choice to diagnose CVST and it will be more accurate when it combined with MRI sequences. This is compatible with Chiewvit *et al.*^[14] who reported that MRV with MRI is reliable as the sole investigation for CVST.

Finally, the aim of this case study is to confirm that MRV in conjunction with MRI can accurately diagnose CVST without risk of radiation exposure, risk of invasive procedures and with no significant limitations. This is compatible with Gustavo *et al.*^[15] who reported that CT venography (CTV) is reliable

imaging modality for detecting cerebral sinus venous thrombosis and it is at least equivalent to MRV, but the risk of radiation exposure, the potential for iodine contrast material allergic reactions, and risk to use it in poor renal function patients are significant limitations. Direct cerebral venography is an invasive procedure, difficult and operator dependent so that it is reserved for cases in which MRV and CTV give inconclusive results and for endovascular therapeutic procedures. Transfontanelle Ultrasound with Doppler may be useful to diagnose CVST only in neonates and young infants.

Conclusion

CVST can be present by a mysterious clinical presentation that makes it as a challenge of diagnosis even by medical imaging by CT and MRI. A combination of MRI and MRV are the best noninvasive imaging modality for diagnosis of CVST without risk of radiation exposure or risk of invasive procedures.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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