Superior Sagittal Sinus Thrombosis Presenting with Hallucinations in the Puerperium: A Case Report

Zylfiqe Hundozi, MD, PhD1*; Nazmie Ibishi, MD, PhD2; Fisnik Jashari, MD, PhD1; Jehona Rrustemi, MD1; Bujar Gjikolli, MD3

1Department of Neurology, University Clinical Center of Kosovo; 2Department of Psychiatry, University Clinical Center of Kosovo; 3Department of Radiology, University Clinical Center of Kosovo

Prishtina, Kosovo

Abstract

Cerebral venous sinus thrombosis is an uncommon cause of stroke presenting with varied presentation patterns. We report a case of a 21-year-old woman with superior sagittal sinus (SSS) thrombosis (SSST) developing after childbirth, presenting with visual hallucinations, severe headache, and tonic-clonic seizures. Time-of-flight magnetic resonance angiography (TOF-MRA) demonstrated the presence of thrombus in SSS. She was treated with low molecular weight heparin (LMWH) followed by warfarin. She had excellent recovery a few weeks after admission and was regularly followed up. Although this condition can be presented with different neurological symptoms, it does not typically present with hallucinations. We suggest that CSVT should be suspected even when a patient presents with an atypical picture in a category of patients at higher risk. (Int J Biomed. 2016; 6(4):294-297.)

Key words: cerebral venous sinus thrombosis ● hallucinations ● tonic-clonic seizure ● puerperium

Introduction

Cerebral venous sinus thrombosis (CVST) is less common than other types of stroke but can be more challenging to diagnose due to its varied presentation patterns; for this reason, it seems to be overlooked not only by general practitioners but also in some specific cases by neurologists as well.[1] CSVT is a multifactorial condition with sex-related specific causes. Among its important etiological factors, pregnancy, puerperium, oral contraceptive use, coagulopathies, intracranial infections, cranial tumors, lumbar puncture, malignancy, dehydration, inflammatory bowel disease, connective tissue disorders, Behcet’s disease, parenetal infections, and various drugs can be implicated.[2] Among other causes, the inherited pro-thrombotic tendencies, such as factor V Leiden mutation, protein S or C and anti-thrombin III deficiencies are also important. However, in 30% of patients the etiology cannot be determined.[3] The clinical signs and symptoms of CVST are relatively nonspecific. The presentation of CVST can include headache, vomiting, papilledema, mental status changes, seizures, and focal neurologic deficit (motor and/or sensory).[4] SSST may present with unilateral paralysis that extends to the other side secondary to extension of the clot into the cerebral veins. Because of the location, this may present as a unilateral lower extremity weakness or paraplegia.[5] Although CVST may present with neuropsychiatric symptoms, it does not typically present with hallucinations.[4]

Because of the very nonspecific and variable clinical picture of CVST, brain imaging plays a crucial role in the diagnosis. The accuracy of magnetic resonance venography (MRV) and TOF-MRA for depiction of cerebral venous thrombosis is better compared to computed tomography angiography (CTA) and MRI.[6]

Treatment options for CVST include anticoagulants, thrombolytic therapy and, in some cases, surgical thrombectomy. The use of heparin and oral anticoagulants is based on a rationale of reversing the causal thrombotic process and of preventing complications.[7] CVST can result in death or permanent disability, but usually has a favorable prognosis.[8]

*Corresponding author: Prof. Zylfiqe Hundozi MD, PhD. Department of Neurology, University Clinical Center of Kosovo, Prishtina, Kosovo. E-mail: zylhundozi@hotmail.com
Case report

A 21-year-old woman presented in the neurologic department complaining of severe headache, speech disturbances, hallucinations and tonic-clonic seizures. She was 4-day postpartum following Caesarean section. She had hypertension and problems with vision (scotomes and blurred vision) during the pregnancy. After antihypertensive therapy with α-methyldopa 250 mg, 4 times daily, the arterial blood pressure was normalized and the vision went to normal. There was no history of epilepsy.

During the hospitalization in the neurologic clinic, a bilateral headache was persistent, and she had hallucinations and two tonic-clonic seizures. In the physical examination, there was no nuchal rigidity but she was dehydrated. There was no neurological deficit. The seizures were controlled with intravenous lorazepam, which was commenced with carbamazepine tablets.

Laboratory blood analysis was performed and the following results were obtained: ESR, 110 mm/hr; RBC count, 4.59 x 10^{12}/L; hemoglobin, 12.3 g/dL; WBC count, 14.7 x 10^{9}/L; hematocrit, 36.4 %; platelets, 206 000; serum glucose level, 6.76 mmol/L; serum creatinine, 0.66 mg/dL. The international normalized ratio (INR) and partial thromboplastin time (PTT) were within normal ranges: 0.9 and 25, respectively. D-Dimer was increased to 752 ng/mL; thrombin time (TT) was 16 seconds. During follow-up, cranial CT and TOF-MRA were obtained, which demonstrated the thrombosed SSS with partial recanalization (Figures 1 and 2).

Discussion

CVST can affect all age groups, particularly women of childbearing age.[9] Pregnancy and puerperium are prothrombotic risk factors. Acute blood loss during delivery, prolonged lying in bed, postpartum infections, sweating, and hyperlipidemia dramatically increase the chance of venous thromboembolism.[10] Cesarean delivery and pregnancy-related hypertension also increase CVST risk.[10,11]

CVST is a rare pathology that presents with a wide spectrum of manifestations. Our patient with cesarean delivery presented with SSST. SSS is most commonly involved with thrombosis. Most patients present with rapid neurological deterioration.[12] The clinical picture is determined by the age of patient, site of CVST, and the presence or absence of parenchymal lesions.[11,13] When a focal brain injury occurs, most common clinical presentations are hemiparesis and aphasia, but other cortical signs and sensory symptoms may be also observed, together with psychosis in rare cases.[14]

Our patient presented with a severe progressive bilateral headache, tonic-clonic seizures, and hallucinations. The headache was bilateral and worsening with sleep. Headaches are the most frequent presenting symptoms (70.6%) of CVST, followed by seizures (47%) and paresis (43%),[11] and have been described as its only clinical presentation in 15% of patients. There is no typical pattern of headaches in CVST. A headache can be of any grade of severity, usually is global and persistent, and has an acute onset.[15]

Intracranial hypertension with local inflammatory reaction determining dilation of vessels in the walls of the affected sinuses is possible, and extravasations of inflammatory proteins could explain the bilateral pain. The pain may also be caused by the stretching of the nerve fibers in the walls of the occluded sinus.[13,15]

There are controversial data on the frequency of seizures after CVST; a prevalence of 10% to 50% has been reported.[16] Most of the seizure types are focal seizures. Rarely, life-threatening generalized tonic clonic seizures can be seen.[17,18]

No clinical trials have studied either the optimal timing or medication choice for anticonvulsant in CVST. Whether to initiate anticonvulsants in all cases of CVST or wait for initial seizures before treatment is controversial. Because seizures
increase the risk of anoxic damage, anticonvulsant treatment after even a single seizure is reasonable.\textsuperscript{14}

In our patient, we made definite diagnosis by MRV, which is the most sensitive diagnostic investigation, but also TOF-MRA has a high accuracy. It has to be underlined that very frequently delays in diagnosis of CVST are common and significant. MRV allows direct visualization of the dural venous sinuses and the large cerebral veins.\textsuperscript{19}

Neuroimaging techniques have allowed a more rapid and accurate diagnosis of these conditions, enabling earlier therapeutic interventions.\textsuperscript{4} Neuroimaging techniques have also demonstrated that the prevalence of CVST is higher than previously reported.

Treatment options for CVST include anticoagulants, thrombolytic therapy and, in some cases, surgical thrombectomy.\textsuperscript{20} Our patient was treated with anticoagulation therapy and had very good recovery. It has been conclusively shown that intravenous heparin is the first-line treatment for CVST because of its efficacy, safety and feasibility. The initial anticoagulation therapy has 3 aims in CVST: a) to prevent thrombus growth, b) to facilitate recanalization, and c) to prevent deep venous thrombosis or pulmonary embolism.\textsuperscript{14}

Post-acute treatment with oral anticoagulants is recommended for up to 6 to 12 months.\textsuperscript{21}

Prognosis of CVST is quite variable, the outcome ranging from total recovery to death. The disease had fatal outcomes during the pre-imaging era, when early diagnosis and effective therapies were not possible, and only supportive care was available. Currently, more clinical studies are reporting a better outcome. Neuropsychiatric manifestation, as in our patient, and pseudotumor cerebri-like presentations carry favorable prognosis while an acute fulminant course, bilateral hemorrhagic infarctions and diffuse cerebral edema are associated with relatively poor outcomes.\textsuperscript{22} Favorable outcomes in obstetric CTSV have been attributed to the assumption that the occlusion is limited and transient with rapid recanalization, or by development of collaterals.\textsuperscript{23} Mortality rates range from 6% to 10%, and independent survival is reported in 90% of patients.\textsuperscript{24}

In conclusion, CVST is a rare pathology that needs a complex diagnostic evaluation. CVST must be considered in young women presenting with any neurologic manifestation related to CNS during puerperium and pregnancy but also when presenting with atypical symptoms, such as hallucinations. TOF-MRA and MRV are the best means of investigation because they make early diagnosis and treatment possible. Generally, correcting the cause can prevent complications.

Competing interests

The authors declare that they have no competing interests.

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