

Case Report

Cerebral Venous Sinus Thrombosis

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Abstract

Cerebral venous sinus thrombosis (CVST) is a rare stroke-like syndrome with significant morbidity and mortality. It primarily affects the young, healthy patient. Recognition of risk factors, symptoms and its treatment can provide full recovery from neurological sequela. We present a case of a young post-partum patient that presented to our Emergency Department with headaches that were thought to be a complication of a post epidural procedure she received during labor and her treatment with epidural blood patches.

Keywords

- Dural sinus thrombosis
- Headache
- Epidural blood patch
- Seizures

ABBREVIATIONS

ED: Emergency Department; CVST: Cerebral Venous Sinus Thrombosis; CSF: Cerebrospinal Fluid; PPD : Post-Partum Day; PMH: Past Medical History; IVF: Intravenous Fluids; CT: Computed Tomography; : MRI/MRV: Magnetic Resonance Imaging with Venography; ICP: Intracranial Pressure

INTRODUCTION

Cerebral venous sinus thrombosis (CVST) is a rare stroke-like syndrome with significant morbidity and mortality that must be kept in every emergency physician's differential diagnosis. This entity is associated with hyper coagulable states and can present with a myriad of signs and symptoms ranging from simple headaches to localizing neurologic deficits, frank seizures, and coma. It primarily affects the young, healthy patient with average age of 39 years old. Prompt recognition of presenting signs and symptoms will lead to the diagnosis and treatment that can prevent significant morbidity and years of life lost [1].

Partum and peripartum periods carry an approximate 10-fold increase in incidence of CVST. Other associated risk factors include infection, genetically acquired pro-thrombotic states, and oral contraceptive use [1]. There are few reported cases of CVST diagnosed after blood patch administration, and none with the acute onset of seizures. We present a case of a CVST occurring in a 2-week post-partum woman who presented with acute, lateralizing neurologic deficits and new-onset seizures immediately following blood patch administration.

CASE PRESENTATION

A previously healthy 33 year-old Pakistani woman at 39 weeks and 3 days gestation was admitted to the obstetrics service in labor after an uneventful pregnancy course. She had an epidural catheter placed which took several attempts and resulted in dural

puncture and cerebral spinal fluid (CSF) leakage. Spontaneous vaginal delivery occurred without complication. The following day, the patient developed a positional headache that was diagnosed as a post-dural puncture headache and treated with an epidural blood patch on post-partum day (PPD) 1. The headache initially resolved, but returned with less intensity the next day. She was discharged home with prescriptions for butalbital-acetaminophen-caffeine and acetaminophen-oxycodone on PPD 2.

On PPD 10, she presented to the Emergency Department (ED) with a gradually intensifying headache which was no longer responding to medication. The headache was described as a constant, dull, severe pressure located in the occiput and the base of neck and was made worse by sitting or standing. She denied any significant past medical history (PMH) or substance abuse disorder. The patient had lived in the United States for several years with her husband and two other children. She denied any recent travel or sick contacts. Physical examination was normal and neurologic examination was non-focal. She received two liters of intravenous fluid (IVF) and butalbital-acetaminophen-caffeine which improved her symptoms. She was discharged with the diagnosis of dehydration and nonspecific headache.

Six days later (PPD 16) she returned to the ED with worsening of her headache. This was so debilitating that she could no longer perform activities such as getting out of bed to taking care of her children. Her headache was now described as global, pounding, and aggravated by changes in position. She denied photophobia, noise or light sensitivity, fever, chills, nausea, or vomiting. Her blood pressure was 126/89 mmHg, heart rate was 98 beats/min, respiratory rate was 18 breaths/min, arterial oxygen saturation was 96% on room-air, and temperature was 98.6 F (37.0 C). She was uncomfortable, clutching her head and crying intermittently. Her physical examination including a neurological exam was otherwise normal.

She was treated with 1 liter of IVF, 4 mg of ondansetron, and 1 dose of butalbital-acetaminophen-caffeine with minimal relief in symptoms. Due to the continued and positional nature of the headache, the anesthesiology service was consulted to evaluate for another blood patch. A second epidural blood patch was performed without complication and this provided moderate resolution of the headache. While the patient was dressing up and preparing for discharge the husband alerted the physician staff that she could not move her leg. Exam showed a new left leg flaccid paralysis. An emergent lumbar MRI was ordered to evaluate for procedural complications. Almost immediately the patient developed left arm tremors with right eye deviation that progressed to a generalized tonic-clonic seizure with urinary incontinence and decelerate posturing. The seizure was terminated with 1 mg intravenous lorazepam. The patient did not recover consciousness and had another seizure two minutes later. At this point rapid sequence intubation was performed to protect her airway. An emergent non-contrast computed tomography (CT) scan of the head revealed a dural sinus thrombosis in the superior sagittal sinus and cortical veins, as well as a small right vertex subarachnoid hemorrhage. Intravenous heparin and fosphenytoin were initiated and the patient was admitted to the Neurosurgical Intensive Care Unit. Warfarin and levetiracetam was started. CT angiography and magnetic resonance imaging with venography (MRI/MRV) confirmed the diagnosis of acute sagittal sinus thrombosis, left transverse sinus thrombosis, and multiple partially thrombosed cortical veins (Figures 1-3). The patient recovered and was extubated the next day. A pro-thrombotic workup was negative including tests for lower extremity deep vein thrombosis, Factor V Leiden mutation, Lupus anticoagulant, Protein C and S resistance, anti-cardiolipin, homocysteine, and pro-thrombin mutation. All neurologic deficits resolved, and the patient had no further seizures. She was discharged home on hospital day 11 on warfarin and levetiracetam. She remained on these medications for 6 months and had complete resolution of the thrombosis confirmed by repeat CT angiography. She continues to have intermittent headaches which are managed by oral medication. She has returned to all of her previous activities.

DISCUSSION

Cerebral venous sinus thrombosis is a rare stroke-like syndrome that can present a diagnostic dilemma to emergency physicians. CVST occurs in about 1 in 100,000 patients per year, has a 3:1 female predominance, and affects the relatively young with an average age of 39 years [1]. It is well known that pregnancy induces a pro-thrombotic state. In fact, thrombosis of any type represents the third leading cause of death in pregnancy [2]. Consequently, the incidence of CVST is increased more than ten-fold to 11.6 in 100,000 during pregnancy and the puerperium. CVST can be a devastating event with greater than 50% mortality if unrecognized. With modern imaging and treatment, 5 to 8% of patients still die within the acute phase and 15% will die or remain dependent at 78 months. Approximately 80% will fully recover if proper and timely treatment is administered [1].

Presentation is widely variable including isolated headache, mood changes, cranial nerve palsies, ocular complaints, aphasia, encephalopathy, TIA/stroke symptoms, seizures or frank coma. Headache is the most consistent presenting complaint and is

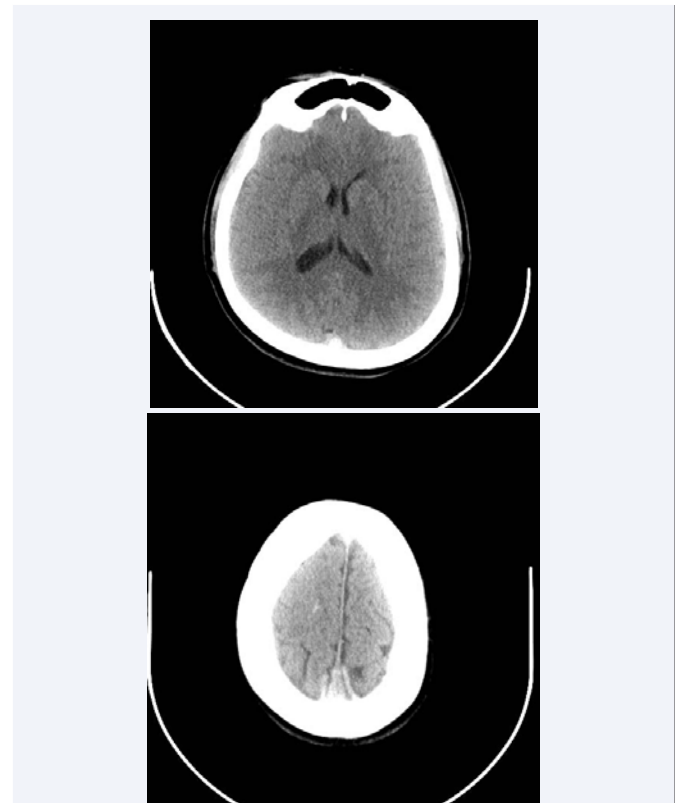


Figure 1 Plain CT scan head with the round shape (A) or dense triangle (B) in the posterior superior sagittal sinus.

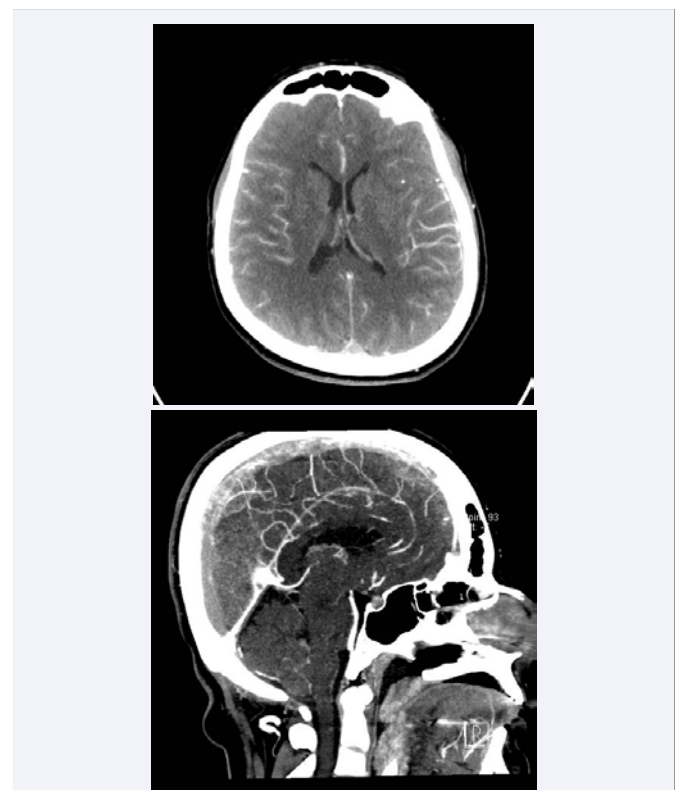


Figure 2 The "Empty Delta Sign" seen in (A) with contrast-enhanced CT scan is a central region lacking contrast in the posterior superior sagittal sinus (B).

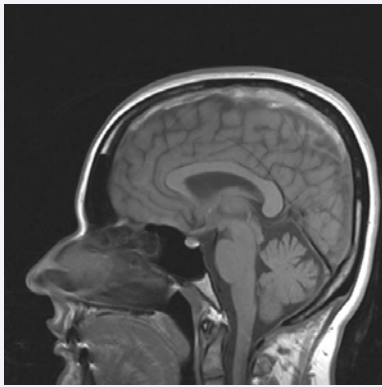


Figure 3 MRV showing a dense area in the superior sagittal sinus.

classically described as slow-onset, dull, localized and worse with recumbency. Some CVST case reports alternatively describe thunder-clap type headaches without the classic positional component. In the most expansive study to date, 37% of patients had some type of focal motor weakness and 39% had seizures on initial presentation [1]. Seizures are more common in the CVST cases with more indolent, chronic presentation and large clot burden. The diversity of symptoms means patients with CVST have the potential to present to any variety of acute, primary care or subspecialist settings. Clinicians must have a high index of suspicion when presented with patients with acute debilitating headaches.

CVST formation results from an identifiable chronic predisposing factor that is coupled with an acute inciting event. Predisposing risk factors include pregnancy, puerperium, genetic or acquired pro-thrombotic conditions, Crohn's disease, sickle cell disease, malignancy, infection, and oral contraceptive use. Inciting events include trauma, jugular catheterization, surgery, infection, dural puncture, and corticosteroid injection. In the largest study to date, 85% of patients had an identifiable cause including 34% with a clear pro-thrombotic condition, 22% with a predisposing genetic condition, and 12% with an acute infection. Only 15% had no identifiable cause [1, 3].

Various imaging techniques are used to confirm the diagnosis with CT angiography and MRI/MRV being most sensitive and specific. Approximately 30% of patients will also have some form of hemorrhage identified on initial imaging [4]. Most are small, low-pressure intra parenchymal hemorrhages of venous origin. Unfortunately, initial non-contrast head CT is normal in nearly 30% of patients with CVST [4]. Nonspecific changes such as focal edema, venous infarction, dilated veins, small ventricles, and subtle diffuse edema may be present. The "Dense Triangle Sign" is a hyperdense triangular or round shape in the posterior superior sagittal sinus that has been described in non-contrast head CT scans (Figure 1). The "Empty Delta Sign" (Figure 2) seen with contrast-enhanced CT scan is a triangular ring of enhancement with a central region lacking contrast in the posterior superior sagittal sinus [4]. Emergency physicians, who strongly suspect CVST should obtain non-contrast CT, followed by contrast enhanced CT, and finally MRI/MRV (Figure 3). There are no specific laboratory tests and nearly 10% of patients with CVST will have a negative D-dimer [5].

Initial treatment is aimed at arresting the thrombotic process, suppressing seizure activity and treating increased intracranial pressure (ICP). Basic measures such as head of bed elevation and airway protection should be followed. Heparin is safe and effective in mortality reduction even in those with small parenchymal hemorrhages. Warfarin should be initiated in the inpatient setting and continued for at least 6 months with goal INR of 2.0-3.0. Rare case reports describe widely variable outcomes with attempted direct thrombolysis and thrombectomy in unstable/refractory patients [6]. Antiepileptic medications can be given for seizures and patients should remain on these for 6 months as well. Inpatient workup should include a thorough evaluation for infection, malignancy, and hypercoagulability including testing for prothrombin gene mutation G20210A, Protein C and S deficiency, Factor V Leiden, lupus anticoagulant, anticardiolipin and anti-beta-2 glycoprotein antibodies [6].

Our case is both unique and typical in several ways. Like many cases before, our patient presented to the ED several times before the final diagnosis was made. Bounce-back patients should always prompt ED physicians to widen their differential diagnosis and workup strategies. The positional features of the headache and presumed 'failed' first blood patch for post-dural puncture headache treatment led to confirmation bias further delaying the final diagnosis. This case is unique in that other than being two weeks post-partum, the extensive pro-thrombotic workup was entirely negative.

This raised the question whether the second blood patch itself might have led to the seizure activity. CVST cases after dural puncture have been described, but most are associated with corticosteroid injection, pre-existing prothrombotic conditions, or infection. Only one has been reported after blood patch placement in which the patient developed an isolated quadrantanopia leading to the CVST diagnosis [7-9]. Dural puncture with CSF leak decreases ICP leading to rostro caudal traction on the cortical sinuses and veins themselves. This traction leads to venous endothelial damage, inflammation and dilation which ultimately causes venous stasis and thrombosis [10-18]. CSF normally drains into the venous sinuses but since these were effectively blocked by thrombosis in our patient, the CSF leak from the dural puncture probably provided an alternative drainage pathway. When the second blood patch was placed, it may have caused a sudden rise in ICP leading to the seizure activity.

CVST is a rare stroke-like syndrome that strikes young, healthy patients and carries significant morbidity and mortality if not identified and treated early. The emergency physician should suspect this in the pregnant or post-partum patient that presents with headache, especially if the diagnosis of post-dural puncture headache is entertained. A high index of suspicion and thorough history should lead the emergency physician to pursue CT imaging and MRI/MRV if necessary. When identified, heparin should be initiated promptly and measures to control ICP and seizure activity should be employed. With prompt identification and treatment this life threatening condition can be easily treated with full recovery for most patients.

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