Cerebral Venous Sinus Thrombosis and Posterior Reversible Encephalopathy Syndrome Coexisting in a Woman: A Rare Coincidence

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Abstract
Cerebral venous sinus thrombosis (CVST) and posterior reversible encephalopathy syndrome (PRES) are two rare diseases which may present with similar symptoms and signs. We report a case with coexisting PRES and CVST in a 34 years old postpartum female presented with multiple episodes of generalized seizures and bilateral vision loss after delivery. MRI brain and venography revealed left transverse sinus, sigmoid sinus and internal cerebral vein thrombosis with vasogenic edema in bilateral parieto-occipital, right temporal and left frontal area, which was suggestive of posterior reversible encephalopathy syndrome (PRES). She was treated with antihypertensive, low molecular weight heparin (LMWH), oral anticoagulant and responded well to the treatment.

Introduction

Posterior reversible encephalopathy syndrome and cerebral venous sinus thrombosis presents with similar kind of symptoms. A clinical diagnosis of PRES includes the presence of headache, seizures, encephalopathy and visual disturbances as well as radiologic findings of focal reversible vasogenic edema best seen on magnetic resonance imaging (MRI) of the brain. The syndrome is most commonly encountered in association with acute hypertension, preeclampsia or eclampsia. The clinical presentation of CVST is similar to PRES, diagnosed best on magnetic resonance venography brain as occlusion of one or more of venous sinus and, or cerebral vein by thrombus. Although both conditions have similar clinical features but, treatment is entirely different; hence we need to differentiate between these two conditions. Here we present a case of a 34 year old female who presented with postpartum eclampsia and diagnosed as a case of PRES with CVST.

Case Presentation

A 34 year old female who was postpartum day one, had multiple episodes of generalized seizures. There was no history of fever, ear or nasal discharge, head injury, neck stiffness or limb weakness. There was no past history of hypertension, diabetes, cerebrovascular accident, epilepsy, oral contraceptive use, any other chronic illness or similar complaints in the past. She also had one full term normal vaginal delivery six years back. There was no past history of preeclampsia or eclampsia. On examination patient was confused, partially responding to verbal commands. Her blood pressure was 180, 110 mm of Hg with pulse rate 90, min and temperature of 99 oF. On cardiovascular and respiratory examination, Per abdomen, uterus was palpable at 16 weeks height and bleeding P/V was minimal. For the control of seizures intravenous magnesium sulphate (1 gm, hr) was given along with antihypertensive drugs (IV Labetalol). Laboratory findings revealed hemoglobin 11.2 gm, dl, total leukocyte count 9000, mm³, platelet count 1.8 lac, mm³. Serum levels of liver enzymes were aspartate transaminase 23 U/L, alanine transaminase 37 U/L, alkaline phosphatase 127 U/L and serum bilirubin levels of 0.7 mg/dl. Renal profile revealed blood urea 32 mg/dl, serum creatinine 1.0 mg/dl, serum uric acid 4.3 mg/dl with serum calcium and phosphate levels 10.6 mg, dl and 3.5 respectively. Serum sodium and potassium levels were 141 mEq/L and 3.6 mEq/L with blood sugar level 126 mg/dl. Urine examination revealed 2+ proteinuria.

On 3rd postpartum day, patient complained of headache and blurring of vision. Fundus examination was done which revealed bilateral papilledema. Plain CT scan brain showed, showing hypodensities in bilateral parietooccipital, right temporal and left frontal region. MRI brain with venography brain (Figure 1, Images A to L) was performed which revealed a hyperintense lesion on T2W, Flair images in bilateral parietooccipital, left temporal and right frontal region which were hypointense on T1W image and shows diffusion restriction on DWI. On ADC maps, hypointensity at right parietooccipital and left occipital region, hyperintense lesions in other areas which were hyperintense on DWI (findings compatible with vasogenic edema) were seen. On MR Venography brain left transverse sinus, sigmoid sinus and cerebral veins were occluded (Figure 2).

Based on the above imaging findings, a diagnosis of PRES (due to uncontrolled hypertension, eclampsia) with CVST (likely due to thrombophilic postpartum state) was made. She was treated with low molecular
She was asymptomatic on follow up after 1 week.

**Discussion**

Cerebral venous sinus thrombosis (CVST) and posterior reversible encephalopathy syndrome (PRES) usually present with similar symptoms and signs such as headache, nausea, vomiting, visual disturbances and seizures. PRES is caused by dysregulation of cerebral blood flow and disruption of blood brain barrier leading to vasogenic edema in brain. The predisposition for posterior circulation in PRES could be due to sparse sympathetic innervation to posterior vasculature. Uncontrolled hypertension (most common risk factor), as in this case, leads to hyperperfusion and cerebral vessel damage due to inadequate autonomic sympathetic response, resulting in interstitial extravasation of proteins and fluids, causing vasogenic edema. MRI is the gold standard for diagnosis and should be performed when PRES is suspected like in this case. Treatment includes aggressive management of hypertension, use of antiepileptic drugs to treat seizures and corticosteroids for vasogenic edema. The prognosis of PRES is excellent if promptly diagnosed and treated.

CVST is the presence of thrombosis in the cerebral veins and, or dural venous sinus that prevents blood from draining out of the brain leading to cerebral ischemia, infarction, hemorrhage, edema, neuronal dysfunction and hyperexcitability. The incidence of CVST is about 3–4 persons per million affecting female more than male. The predisposing factors to CVST are genetic (factor V Leiden, protein C, S deficiency, hyperhomocysteinemia,
antithrombin III deficiency, prothrombin gene mutations etc) and acquired prothrombotic states (pregnancy, postpartum, antiphospholipid antibody syndrome, oral contraceptive use, hormonal replacement therapy), polycythemia, sickle cell disease, thrombocytopenia, paranasal sinus infection, otitis media, dehydration, uncontrolled diabetes, thyroid dysfunction, connective tissue disease, vasculitis, inflammatory bowel disease, liver disease, smoking head injury, folic acid and vitamin B12 deficiency (acquired hyperhomocysteinemia) etc. A clinical diagnosis of CVST should be made on the basis of clinical presentation and in the presence of predisposing factors for venous thrombosis. The most sensitive technique for the diagnosis of CVST is magnetic resonance venography seen as occlusion of one or more venous sinus and, or cerebral vein. Other findings like cerebral ischemia, infarction with diffusion restriction on DWI and diminution of ADC values (suggestive of cytotoxic edema) and vasogenic edema similar to PRES can be seen. Treatment includes identification and elimination of the underlying cause, management of raised intracranial pressure and anticoagulation, initially with low molecular weight heparin, after three to four days warfarin is started and both overlapped for two to three days then heparin is withdraw. Anticoagulants are usually given for a period of six months after a first episode or longer when there is persistence of predisposing factors. Other treatment modalities in the form of endovascular thrombolysis and mechanical thromboaspiration alone or in combination can be tried in patients with poor prognosis.6

Here we have presented a case of 34 years old female with postpartum eclampsia having PRES with CVST, although two different clinical entities, with different etiopathogenesis and treatment, but similar clinical and neuroimaging features. Two common complications in postpartum period, CVST and PRES coexisting in one patient as in our case is a rare coincidence.

Conclusion

PRES and CVST are two diseases which have similar manifestations but different treatment modalities. These two diseases can have similar clinical and neuroimaging features involving posterior circulation, also clinical setting to develop venous thrombosis and reversible encephalopathy. These should be borne in mind while evaluating postpartum headache, vision loss and, or seizures. Early diagnosis and treatment of PRES and CVST have excellent outcomes and prognosis.

References