

Cerebral Venous Sinus Thrombosis and Posterior Reversible Encephalopathy Syndrome in a Preeclamptic Woman

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ABSTRACT

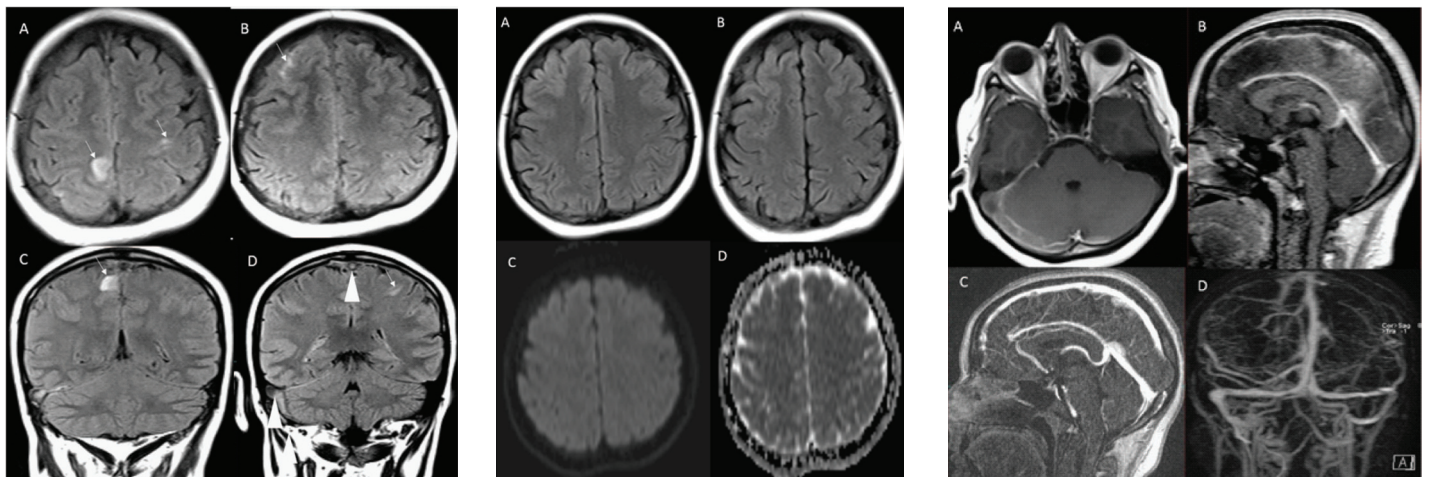
Cerebral venous sinus thrombosis (CVST) and posterior reversible encephalopathy syndrome (PRES) are two rare diseases which may present with similar symptoms and signs. We report a case with coexisting PRES and CVST in a preeclamptic woman. A 24-year-old woman, G₂ P₁, at 33 wk and 5 d of pregnancy presented with headache. Her blood pressure was 180/120 mmHg and urinary test revealed +3proteinuria. Cesarean section was performed with indications of severe preeclampsia, intrauterine growth retardation and fetal distress. Cranial MR venography of the patient revealed thrombi in superior sagittal sinus, confluenssinuum, right transverse and right sigmoid sinus and diffusion MRI showed increased signal intensity (vasogenicoedema) in cortical and subcortical areas of parietooccipital and posterior frontal lobes. The patient was treated with magnesium sulfate and heparin successfully. After treatment period, control cranial MRI and diffusion MRI were normal. Thrombi within the sinuses were totally regressed. Albeit rare, these two diseases should be kept in mind especially in preeclamptic/eclamptic patients that present with neurological symptom and/or sign.

Keywords: Cranial MRI, Neurological symptoms, Pregnancy, Sinus thrombosis

CASE REPORT

A 24-year-old woman, G₂ P₁, at 33 wk and 5 d of pregnancy, presented with headache for two days' duration. She had a history of normal vaginal delivery two years back and she had no history of oral contraceptive use. At presentation her blood pressure, heart rate and body temperature were 180/120 mmHg, 88 bpm, and 36.8°C, respectively. She was hospitalized in our clinic for evaluation of severe pre-eclampsia. Cervical dilatation and effacement were not present on vaginal examination. Ultrasonographic examination detected a growth restricted fetus with fetal biometric measurements as follows: biparietal diameter 32 wk, abdominal circumference 30 wk and femur length 32 wk of gestation and Doppler examination revealed an absent end-diastolic velocity in the umbilical artery of the fetus. Her laboratory findings were as follows: aspartate transaminase 28 U/L; alanine transaminase 8 U/L; lactate dehydrogenase 299 U/L; hematocrit 35.3%; platelets

346,000/mm³; and 3+ proteinuria in spot urine sample. Two doses of Betamethasone 6 mg was administered intramuscularly for fetal lung maturation. Antihypertensive treatment with nifedipine 10 mg was started. Fetal heart rate monitoring showed absence of fetal heart rate variability. A female baby weighing 1760 g (Apgar 7/9) was delivered by cesarean section with indications of severe preeclampsia, intrauterine growth retardation and fetal distress. Prophylactic intravenous MgSO₄ (1g/h) treatment was started due to prodromal symptoms of headache and visual disturbance and continued 24 h after delivery. Hematocrit was 35.3%; platelet count was 346,000/mm³ on postoperative day 1. On postoperative day 2, patient was consulted with Neurology Clinic since patient's headache worsened and she had loss of vision. On neurologic examination, right homonymous hemianopia and papilloedema were detected. On noncontrast cranial computed tomography, hyperdense signal appeared in superior sagittal sinus and right lateral sinus due to



[Table/Fig-1]: FLAIR (A-D) images show gyriform hyperintense signal changes at cortico subcortical areas of bilateral posterior superior frontal, and parasagittal area of right frontoparietal region compatible with PRES in a pregnant patient with cerebral venous thrombosis before treatment (arrows). Besides coronal FLAIR (D) image demonstrate thrombosis at superior sagittal sinus and sigmoid sinus (arrowheads). **[Table/Fig-2]:** After treatment, axial FLAIR (A,B) and diffusion weighted (C,D) images were normal **[Table/Fig-3]:** On Contrast enhanced T1 weighted (A,B), There is thrombus filling the lumen of superior sagittal and sigmoid sinus. After treatment contrast enhanced sagittal T1 weighted (C) and MIP images (D) show the normal cerebral sinuses

slow flow insinuses. Cranial MRI, diffusion weighted MRI (DWI) and MR venography was performed. There were gyral hyperintense signal changes at cortical-subcortical areas of bilateral posterior superior frontal and parasagittal areas of right frontoparietal region in the FLAIR weighted images. In diffusion weighted images (DWI), there were b: 1000 hyperintense, ADC map hypointense (diffusion restriction) at parasagittal space of right frontoparietal region but there were no restriction at other areas and there were signal changes in diffusion compatible with vasogenic oedema. These findings were consistent with PRES [Table/Fig-1a-d]. There were thrombosis in superior sagittal sinus, confluens sinuum, right transverse and right sigmoid sinus in MR venography and MRI. There was contrast enhancement on sinus walls and absence of flow within sinuses in contrast images which is designated as empty delta sign.

She was admitted to neurology intensive care unit. Anticoagulation with low molecular weight heparin (LMWH) was started and patient was discharged from hospital on postoperative day 6. LMWH was continued for one month and then warfarin treatment was planned for six months. In order to investigate predisposing factors underlying the disease, factor V Leiden mutation, prothrombin polymorphism, Antithrombin III levels, protein C and S levels, homocysteine levels, antinuclear antibody, Anti-Cardiolipin IgM antibody were studied. All laboratory tests were within normal range. After treatment period, control cranial MRI and DWI were normal [Table/Fig-2a-d]. Thrombi within the sinuses were totally regressed [Table/Fig-3a-d].

DISCUSSION

Cerebral venous sinus thrombosis (CVST) is a rare disease with an annual incidence of 3 to 4 cases per one million in which thrombolytic processes occur in cerebral venous system [1]. Predisposing factors for CVST consist of pregnancy, puerperium, oral contraceptive use, coagulation disorders (acquired or hereditary), systemic diseases (e.g.; vasculitis such as Behçet syndrome or systemic lupus erythematosus), malignancies and infective causes [2]. Since there is a tendency for thrombosis in pregnancy and postpartum period, the disease is thus most commonly encountered during pregnancy.

Superior sagittal sinus and lateral sinuses are frequently affected sinuses [3]. Mostly, more than one sinus is occluded in 50% of CVST patients [4]. Clinical presentation is widely variable. Onset of the disease may be acute, subacute, or chronic. Although headache is the most common presenting symptom, patients also present with focal neurologic deficits, intracranial hemorrhage, convulsions, altered consciousness and papilloedema [3,5]. Symptoms and signs change not only according to sinuses that are involved but also to progression of the disease. The diagnosis of CVST can be arduous due to lack of specific signs and symptoms of the disease.

Although the diagnosis of the disease is not easy due to wide variety of symptoms and signs, it should be considered in differential diagnosis of patients that present with any neurological symptom and/or sign. Differential diagnosis of postpartum headache includes migraine headache, spinal headache, preeclampsia/eclampsia, CVST, posterior reversible encephalopathy syndrome (PRES), stroke, subarachnoid hemorrhage, meningitis, and cerebral arterial thrombosis [6]. Initial diagnostic work-up should encompass past medical history, duration of headache, associated symptoms and or signs, and response to antihypertensives and analgesics. Neuroimaging should not be omitted in the management algorithm of patients with headache complaint postpartum and should be scheduled in patients with neurological deficit and cases not responding medical treatment [6].

PRES is a condition which presents with headache, seizures, and visual disturbance and the disease is mostly associated with severe hypertension and eclampsia [7]. Suggested underlying mechanism

of the disease is the disruption of autoregulation system of the brain due to hypertension which results in vasogenic oedema in especially occipital and parietal regions of the brain [8]. Autoimmune conditions may also take part in the pathophysiology leading to endothelial dysfunction [9]. Clinical findings resemble presentations of CVST and headache is the most common symptom in CVST patients whereas seizure is the most common clinical presentation of PRES [5,9]. Early diagnosis of PRES is important in order to prevent secondary complications such as status epilepticus, intracranial hemorrhage and ischemic infarction [9].

MRI is the gold standard in the diagnosis and should be performed when PRES is suspected. The lesions appear bright on T2 weighted and FLAIR images. FLAIR images are more sensitive than T2 alone in diagnosis of PRES. The typical lesion is vasogenic oedema predominantly in the occipital lobe, which may be patchy or confluent; but almost all patients (90–98%) have oedema in the parietooccipital region [10]. Typical PRES lesions on MRI represent vasogenic oedema which can be distinguished by DWI. DWI sequences enable the measurement of movement of water molecules. Restricted diffusion, such as in the case of a cytotoxic oedema, is characterized by hyperintensity in b: 1000 images and hypointensity in ADC maps. In case of vasogenic oedema there is an increase in diffusion. DWI shows an iso or hypointense signal whereas ADC maps show increased signal intensity. In our case there were signal changes in diffusion compatible with vasogenic oedema that was totally regressed in control.

In treatment of PRES, Mg_{SO₄} is used for anticonvulsant effect and also in order to decrease vasogenic oedema [7]. Antihypertensive medications may be prescribed in severe hypertension to control blood pressure. Optimal treatment of CVST is anticoagulation with intravenous unfractionated heparin or subcutaneous low molecular weight heparin [3]. Thrombolytic treatment may be an alternative in cases unresponsive to heparin anticoagulation. Cases in which pregnancy and puerperium are underlying factors, prognosis is generally good [5].

CVST and PRES are two conditions that have corresponding clinical presentations. To make differential diagnosis of these diseases, neuroimaging (especially FLAIR images and DWI) should be performed in patients that present with neurological symptoms.

CONCLUSION

We presented a pre-eclamptic woman with coexisting PRES and CVST that was treated with Mg_{SO₄} and heparin successfully and recovered totally, in which our management might be useful for future cases presenting with this unusual diagnosis. Although signs and symptoms of two diseases are similar, treatment modalities are different. These two diseases should be kept in mind especially in preeclamptic/eclamptic patients not only by obstetricians, but also by radiologists.

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