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*Case Report*

# Neurologic Syndromes in Post-partum Women: Posterior Reversible Encephalopathy Syndrome (PRES) vs. Reversible Cerebral Vasoconstriction Syndrome (RCVS), 2 Sides of One Coin?

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## Abstract

Posterior Reversible Encephalopathy Syndrome (PRES) and Reversible Cerebral Vasoconstriction Syndrome (RCVS) constitute a spectrum of disorders characterized by acute-onset headache, altered sensorium, hypertension, visual field defect and seizures, with radiological features of vasogenic edema particularly in parieto-occipital regions of brain. In PRES, the typical feature is vasogenic cerebral edema which is mostly reversible, while in RCVS, cytotoxic edema may persue resulting from multifocal vasoconstriction, which is also reversible with treatment producing complete recovery. Important causes of PRES are: Hypertension secondary to Preeclampsia/Eclampsia, Infections with or without CNS involvement, may be bacterial or viral, occasionally Herpes simplex encephalitis. Autoimmune disease/SLE. Here we report a woman without history of pregnancy induced hypertension who developed progressive hypertension early post- partum and presented with features of both PRES and RCVS. Early recognition of her illness with clinical and radiological features followed by appropriate treatment led to prompt response and prevented a potential threat to her life.

**Keywords**: Reversible encephalopathy, Vasogenic edema, Reversible vasoconstriction, Ischemic infarct, Nimodipine

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## INTRODUCTION

Posterior Reversible Encephalopathy Syndrome (PRES) constitutes a spectrum of disorders characterized by acute-onset headache, altered sensorium, visual field defect and seizures, with radiological features of brain edema particularly in parieto-occipital regions [1,2]. PRES is not a disease or diagnosis by itself, but a condition conglomerating a group of disorders of different etiology with a distinct pathogenesis and characteristic neurological features.

Important causes of PRES are:

- Hypertension secondary to Preeclampsia/Eclampsia,
- Infections /Sepsis with or without CNS involvement, may be bacterial or rarely, viral, Autoimmune disease/SLE./ cancer chemotherapy.

- Essential hypertension with Encephalopathy [1,3].
- In Reversible Cerebral Vasoconstriction Syndrome (RCVS), cytotoxic edema with endothelial dysfunction may persue from multifocal vasoconstriction in cerebral arteries which is also reversible with treatment producing complete recovery.

## CASE PRESENTATION

A 36 yrs. old woman, para 1, living 1 underwent an LUCS and delivered a normal baby. She didn't have pregnancy induced hypertension or PET during her pregnancy, as her blood pressure was normal, there was no edema and urine examination did not show proteinuria. Her post-operative recovery was uneventful and she was discharged after 4 days. 2 days after reaching home she noticed swelling of her legs and face with headache. Her blood pressure was 150/90 mm Hg. She was treated with Amlodipine. Headache persisted and after another 4 days she developed blurring of vision and weakness of all 4 limbs. There was no seizure or any abnormal movement. Her blood pressure examined by her family physician was 170/100. Her first pregnancy and labour was uneventful.

She was referred to and admitted in our hospital. On initial evaluation she was having edema in both legs, pallor was absent. Her temperature was normal. Blood pressure was 175/100. There was no respiratory distress. Neurologically she was conscious and oriented to her surroundings, her speech was normal. Her pupils were normally reacting to light and accommodation. There was no RAPD. A field defect was present as left homonymous hemianopia. Fundoscopy was normal. Motor system examination revealed decreased power in all 4 limbs (MRC grade 4/5) with hypertonia and exaggerated knee jerks on left side. Plantar response was extensor in left side.

After admission she was given an intravenous infusion of labetalol which reduced her initial blood pressure to 150/90 mm Hg. Telmisartan 40 mg daily was started to control her blood pressure thereafter. MRI brain revealed bilateral hyperintense lesions in parieto-occipital and frontal cortex and watershed areas in T2 and flair images and diffusion restricted lesions in DW images (Figures 1-5). Her MR angiogram revealed multifocal vasoconstriction in cerebral arteries with diffuse beaded appearance (Figure 6). MR venogram was normal (Figure 7). EEG showed periodic bihemisheric (Rt>Lt) sharp and slow waves (Figure 2a). Her urinalysis showed mild proteinuria, sterile on culture. CBC, ESR, CRP, blood sugar, liver and kidney functions were normal.

Antinuclear antibody was negative. Her ECG, echo cardiogram including TEE did not show any significant abnormality, indicating absence of PFO or any other source of embolism.

She was diagnosed as RCVS and/or PRES disorder superimposed on post-partum hypertension. Oral nimodipine was added to control intracranial vasospasm. Levetiracetam was added as her EEG showed bihemisheric sharp waves, though she did not have any seizure episode. As her blood pressure settled down, she regained power of her limbs and normal vision. She was discharged after another 7 days of treatment. Physiotherapy was instituted.

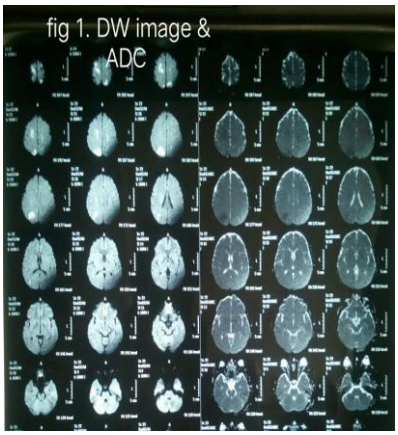


**Figure 1.** DWC image and ADC.

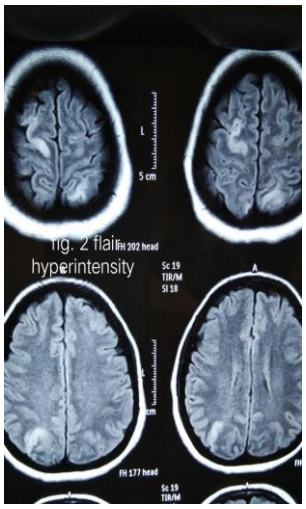


**Figure 2.** Flair hyperintensity.

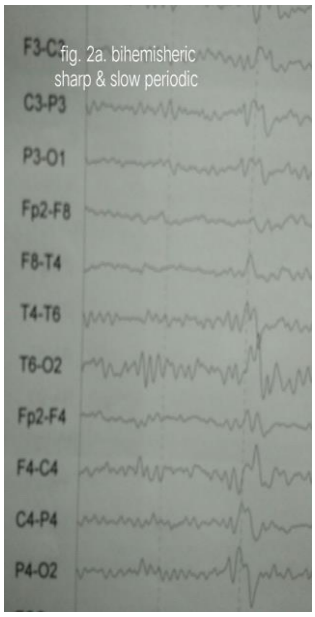


**Figure 2a.** Bihemispheric sharp and slow periodic.

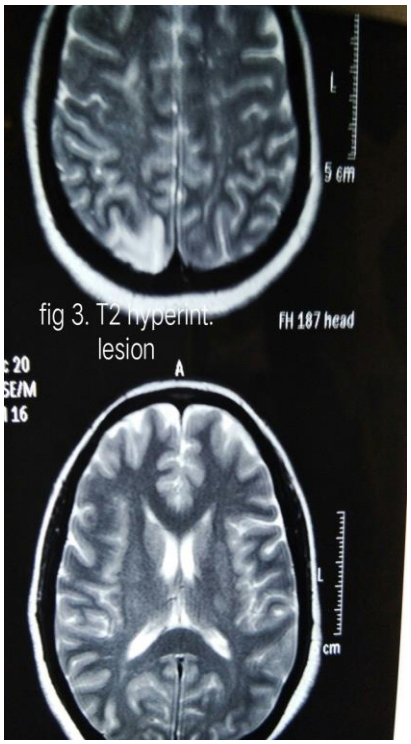


**Figure 3.** T2 hyperintense lesion.

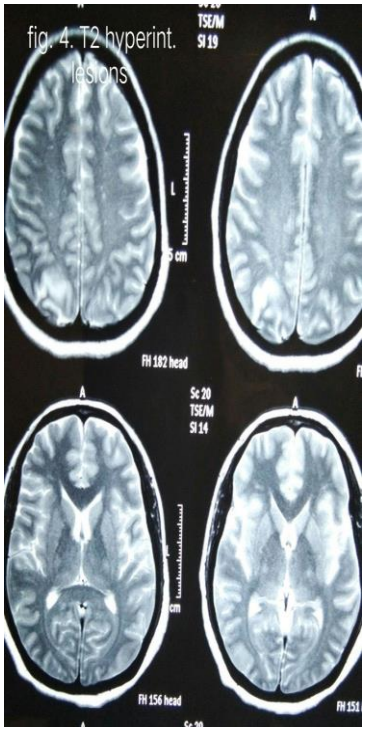


**Figure 4.** T2 hyperintense lesion.

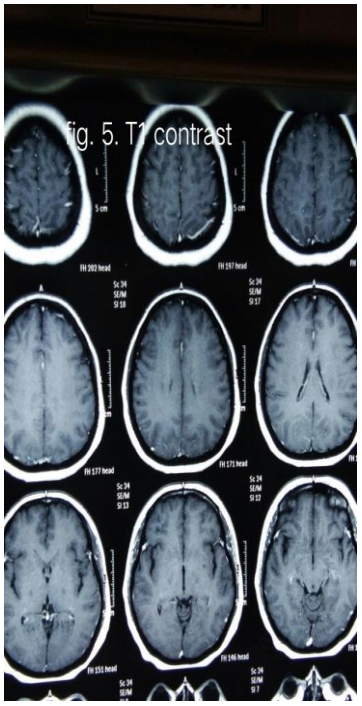


**Figure 5.** T1 contrast.

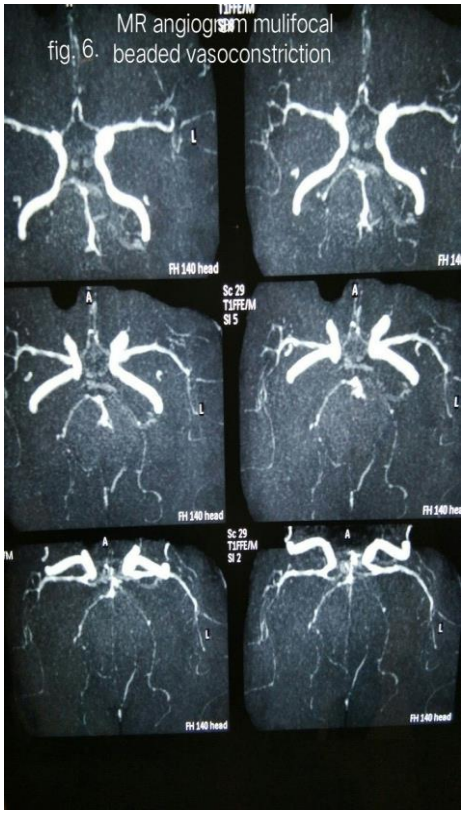


**Figure 6.** MR angiogram multifocal beaded vasoconstriction.

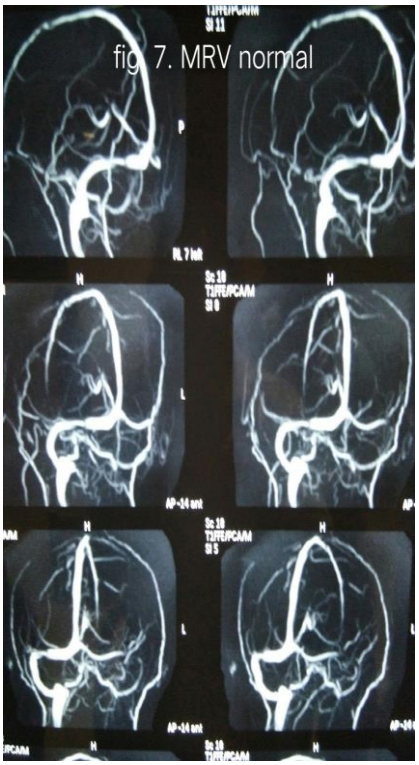


**Figure 7.** MRV normal.

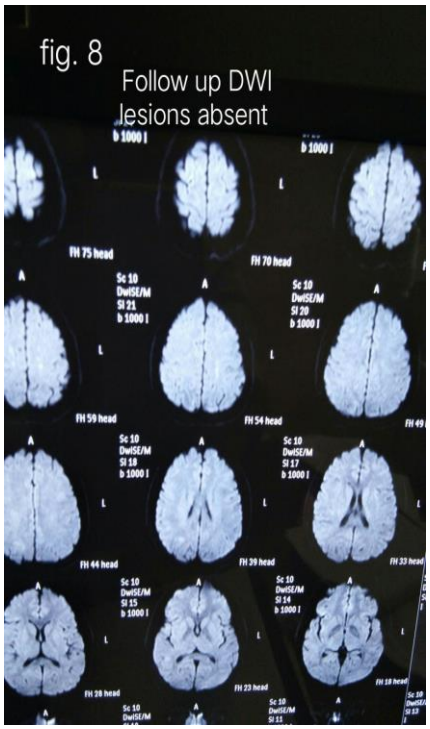


**Figure 8.** Follow up DWI lesions absent.

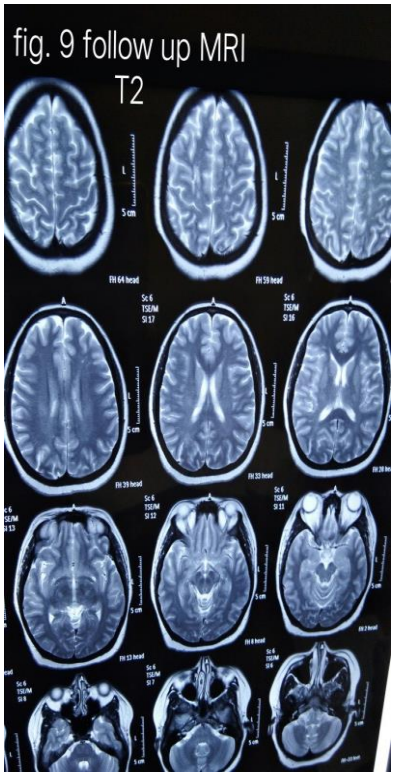


**Figure 9.** Follow up MRI T2.

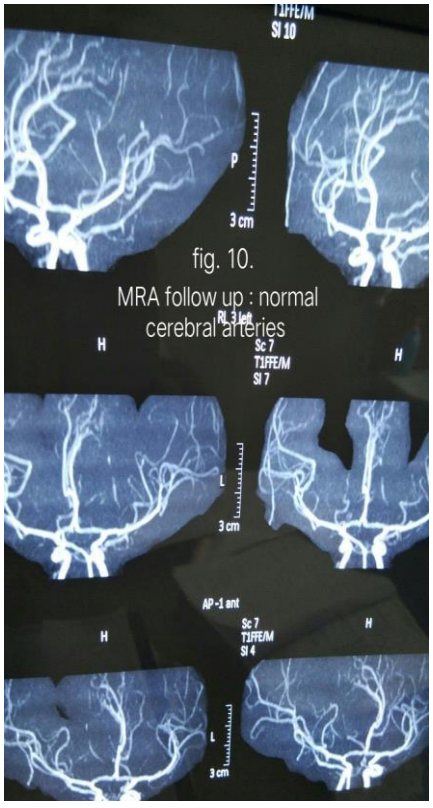


**Figure 10.** MRA follow up: Normal cerebral arteries.

She was advised to continue antihypertensive treatment with Telmisertan and nimodipine with regular blood pressure check for a month and have a repeat MR angiogram thereafter.

Her follow up brain MRI revealed disappearance of T2WI, flair and DWI lesions (Figures 8 and 9).

Repeat MR angiograms (Figure 10) after 1 month showed normal caliber of cerebral arteries with disappearance of vasoconstriction. Clinically her powers of limbs were normal with normal tone and reflexes.

## DISCUSSION

Both PRES and RCVS constitute a spectrum of disorders characterized by acute-onset headache, altered sensorium and visual impairment [1,2]. Seizures may occur, with radiological features of brain edema particularly in parieto-occipital regions, and /or multifocal vasoconstriction in cerebral arteries with endothelial dysfunction and ischemic infarcts [3].

PRES/RCVS are not a disease or diagnosis by itself, but a condition conglomerating a group of disorders of different etiology with a distinct pathogenesis and characteristic neurological features.

It gives neurologists the opportunity for prognostic analysis of the index disorder and adopting measures to prevent clinical deterioration.

**PATHOGENESIS**

Exact pathogenesis of PRES is not known, but acute hypertension with failure of cerebral autoregulation causing breakdown of blood brain barrier is the presumed mechanism of cerebral vasogenic edema [1,4].

The relative lack of sympathetic innervation in the posterior circulation is postulated to inflict parieto-occipital part of the brain, though occasionally anterior brain like insular and fronto temporal area can be affected.

Another theory suggests systemic inflammatory states causing endothelial dysfunction as the cause of PRES. This theory is supported by the common association between PRES and systemic inflammatory conditions such as sepsis, preeclampsia, transplantation and auto immune diseases.

Alternatively, some investigators have proposed vasospasm as the cause of reversible edema that progresses to cytotoxic edema and infarction. This theory is also the basis of Reversible Cerebral Vasoconstriction Syndrome (RCVS).

Thus PRES and RCVS can be considered as spectrum of disorders with similar pathogenesis and manifestations [3,4].

## DIAGNOSIS AND TREATMENT

Brain imaging is the cornerstone in confirming a diagnosis of PRES. Although vasogenic edema can be visualized on non-contrast Computed Tomography (CT) in some patients, brain MRI, especially the T2-weighted and Fluid Attenuated Inversion Recovery (FLAIR) sequences are much more sensitive depicting localised areas of edema. As soon as the condition is postulated, treatment should be provided with appropriate antihypertensives and supportive measures to control brain edema. Initial reduction of arterial pressure should not exceed 20% as further lowering is likely to induce cerebral ischemia.

RCVS with multifocal vasoconstriction should be treated with nimodipine and other supportive measures. Antibiotics should be given to patients susceptible to sepsis.

Our patient was normal till her delivery, and presented with progressive hypertension after delivery with swelling of legs and proteinuria indicated post- partum hypertension. Brain MRI and MRA revealed multifocal vasoconstriction with infarcts/ vasogenic edema which subsided after treatment ensuing complete clinical and radiological recovery thus justifying RCVS and/or PRES.

## CONCLUSION

Postpartum hypertension can be of serious consequences, with rapid progression of vasoconstriction, brain edema and ischemic cerebral infarction. Clinicians need to be aware of the potential complications of this condition. Postpartum women with acute neurologic symptoms require prompt investigation with non-invasive cerebrovascular imaging and close monitoring for possible secondary deterioration.

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