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Case Report

Reversible cerebral vasoconstriction syndrome in a young primigravida woman with pre-eclampsia

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ABSTRACT

Reversible cerebral vasoconstriction syndrome (RCVS) is characterised by severe headache and is associated with reversible segmental vasoconstriction of cerebral arteries. Conditions associated with RCVS are commonly pregnancy with or without pre-eclampsia, neurological procedures, head trauma. Thunderclap headache is the chief clinical presentation. Visual disturbances and focal neurological deficits are also frequently encountered. Posterior reversible encephalopathy syndrome and RCVS are often overlapping and hence most cases of RCVS are diagnosed late. We reported a young primigravida who had no comorbidities presenting to the ER with elevated blood pressure and generalised tonic and clonic seizures. Post-delivery her headache persisted and clinically her neurological status started deteriorating. Later she was diagnosed as RCVS. Treatment is based on expert opinion. Nimodipine, nifedipine or verapamil have been used in most patients.

Keywords: Reversible vasoconstriction syndrome, Posterior reversible vasoconstriction syndrome, Thunderclap headache

INTRODUCTION

Reversible vasoconstriction syndrome (RCVS) also known as isolated benign cerebral vasculitis is characterised by severe headache, reversible segmental vasoconstriction of cerebral arteries and occasionally complicated by ischemic or haemorrhagic stroke. RCVS affects people aged between 20-50 years. RCVS is commonly associated in pregnancy, neurological procedures, head trauma. Thunderclap headache is the chief clinical presentation. Other symptoms associated with RCVS include vomiting, photophobia, phonophobia, visual changes, hemiplegia, ataxia, dysarthria, aphasia, and seizures.[1] These deficits include visual disturbances, photophobia, blindness, focal facial or limb weakness, dysarthria, and ataxia. Generalised tonic-clonic seizures occurred in 17%.[1] Most significantly, severe and permanent neurological deficits and even death may occur as a consequence.

Incidence of RCVS is 3 per one million adults. RCVS affects females more commonly, with female to male ratios- 2:1 to 10:1.[2] The frequency of focal neurologic deficits ranged from 9 to 63 percent.[2] Hemiplegia, tremor, hyperreflexia, ataxia, and aphasia can develop. Visual deficits, including scotomas, blurring, hemianopia, and cortical blindness, are common, and these patients typically have concomitant Posterior reversible leukoencephalopathy syndrome (PRES). Magnetic resonance imaging (MRI) of the brain is normal in over 50 percent of patients with RCVS. Infarcts are often bilateral and symmetrical, located in arterial watershed (i.e.; border-zone) regions of the cerebral hemispheres or in the cortical-subcortical junction.[3]

The pathogenesis of RCVS is poorly understood. It may occur spontaneously (primary RCVS) or be triggered by endogenous or exogenous substances (secondary RCVS) such as cannabis, nicotine, SSRI, predisposing conditions

such as pregnancy, pre-eclampsia, hypercalcemia.[3] There is an interaction between sympathetic overactivity and endothelial dysfunction, hence causing a break in autoregulatory mechanisms.[4] It is postulated that the mediators of vasospasm like endothelin-1, serotonin, nitric oxide, different mechanisms.

Treatment is based on expert opinion. Nimodipine, nifedipine or verapamil have been used in most patients.

CASE REPORT

A 19 year-old primigravida at 36 weeks of gestation presented to the ER with two episodes of GTCS at home . She was received at the ER in post-ictal state, conscious yet disoriented. On receiving her was BP recorded as 160/100 mmHg. On examination pedal edema was noted, her uterus corresponded to 36 weeks and was relaxed and not tense or tender and Fetal heart rate was good. Lab investigations were drawn. Injection labetalol 40 mg IV was given and injection magnesium sulphate 4 g loading dose was given and patient was shifted to OR for emergency lower segment caesarean section- she delivered a 2.3 kg live baby with APGAR 7/10, 8/10. Intra-operatively mild atoncity of uterus was noted and managed medically. Post operatively maintenance dose of magnesium sulphate was restarted. Laboratory investigations revealed hemoglobin 10.6 g/dl, platelets 1.27 lakhs/mm^3, uric acid 12.1 mg/dl, serum creatinine 1.11 mg/dl. On post-operative day 1, her serum creatinine was elevated- 1.5 mg/dl- diagnosed as acute kidney injury and correction was given, subsequent results were normal. Ophthalmic evaluation showed grade 1 hypertensive retinopathy.

On post-operative day 1 patient was transfused with one unit of packed cell in view of hemoglobin 8.1 g/dl. Patient complained of persistent headache and on examination was drowsy. MRI was done and showed features of PRES with no significant occlusion of cerebral, vertebral and basilar arteries. EEG showed continuous epileptiform activity and hence leviciteram injection was started. However, patient continued to be irritable and agitated neurology review sought and patient developed complex partial seizure during the review.

RCVS was suspected and patient was shifted to ICU in view of persistent epileptiform activity and decreased mentation. Her blood pressure was persistently elevated and patient was started on tab nimodipine 30 mf BD, injection labetalol infusion and injection fosphenytoin. CT angiogram of head and neck revealed mild- moderate narrowing of cavernous and supra-clinoid segments of ICA, MCA and ACA suggestive of RCVS. Her blood pressures were continued to persistently be elevated and antihypertensive medications were escalated to tab clonidine 100 mcg TDS and tab prazosin 5 mg, tab nifedipine- sustained release was added. On post-op day 5- lumbar puncture was done which showed normal study. Gradually the patient improved during the course of stay, she was oriented and was able to follow commands.

Injection labetalol infusion and 3% NaCl transfusion tapered and stopped and BP control was achieved. Repeat EEG showed no epileptiform activity, CT brain showed resolving white matter edema in parietal, occipital and temporal lobes with now new lesion. Patient was discharged with multiple antihypertensive medications, which were gradually tapered over weeks. At present she is on tab nifedipine and is performing routine day to day house hold activities.

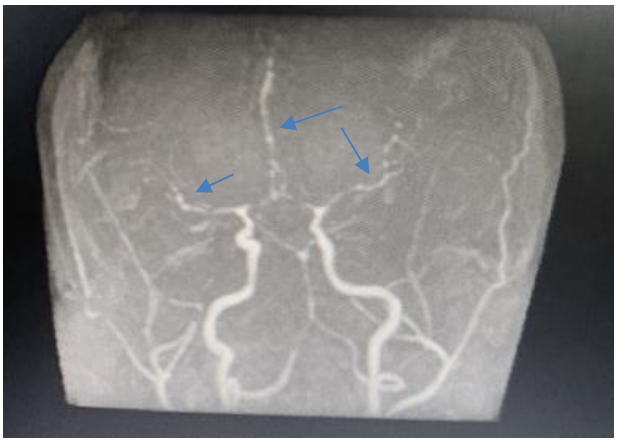

Figure 1: string of bead appearance.

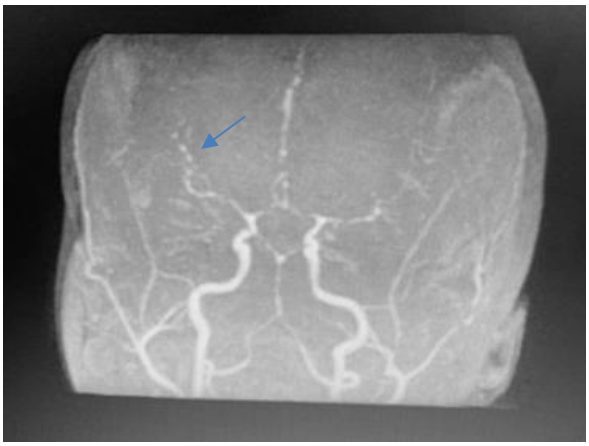

Figure 2: Narrowing of MCA.

DISCUSSION

RCVS affects females more commonly. RCVS is associated with pregnancy, migraine use of vasoconstrictive drugs, neurosurgical procedures, hypercalcemia, cerebral venous thrombosis. Hemiplegia, tremor, hyperreflexia, ataxia, and aphasia develop. Visual deficits, including scotomas, blurring, hemianopia, and cortical blindness, are common, and these patients typically have concomitant PRES. MRI of the brain is normal in over 50 percent of patients with RCVS. Infarcts are often bilateral and symmetrical, located in arterial

watershed (i.e.; border-zone) regions of the cerebral hemispheres or in the cortical-subcortical junction. Routine blood tests, inflammatory markers, and cerebrospinal fluid analysis are typically normal in RCVS. CTA and MRA can identify seventy percent of cases by revealing diffuse reversible cerebral vasoconstriction that appear as 'string of beads' on angiography with complete resolution within 1-3 months. Initial MRI is normal during the first week in 30-70% of cases; 10% of patients have MRI abnormalities consistent with PRES.

Currently, there is no approved treatment for RCVS. Any potential drugs or triggers should be discontinued or avoided in secondary RCVS. Verapamil, nimodipine, and other calcium channel blockers may help reduce the intensity and frequency of the headaches. Intra-arterial verapamil has been shown to improve radiological vasospasm, but improvement in clinical outcomes remains to be proven.[6] The rate of permanent neurological disability is surprisingly low.

CONCLUSION

RCVS is a clinical entity and neurological emergency that is being diagnosed with increasing frequency but is still under recognized, and a high index of suspicion is essential.

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