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Recanalization of Post-Partum Reversible Vasoconstriction Syndrome with Cilostazol Treatment

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ABSTRACT

Reversible cerebral vasoconstriction syndrome (RCVS) is a coined term to express conditions that manifest narrowing of the cerebral arteries. The most common manifestation of a patient with RVCS is severe headache that is thunderclap in presentation, however, some may present with focal neurologic deficits related to the site of edema or vascular occlusion and even cortical manifestation such as seizure. The current treatment for RCVS is still observational, which includes use of calcium channel blockers and intra-arterial vasodilation. In this case report, Cilostazol, a phosphodiesterase 3 inhibitor with a myosin light chain kinase enzyme inhibitor, can be used as a treatment for recanalization of constricted cerebral arteries.

Keywords: RCVS, Cilostazol, Post-partum

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Introduction

Reversible cerebral vasoconstriction syndrome (RCVS) represents a group of conditions that show reversible multifocal narrowing of the cerebral arteries with clinical manifestations that typically include thunderclap headache and sometimes include neurologic deficits related to brain edema, stroke, or seizure. The clinical outcome is usually benign, although major strokes can result in severe disability and death in a minority.[1]

The pathophysiology of the abrupt-onset headache and of the prolonged but reversible vasoconstriction is not known. The common manifestation is headache similar to subarachnoid hemorrhage. Thunderclap in presentation with or without neurologic deficit.

In adults, RCVS predominantly affects women, with female to male ratios ranging from 2:1 to 10:1, depending on the case series. [2]

The mean age of affected individuals across published studies is 42 to 44 years, with an age range of 4 months to 65 years [3,4] RCVS occurs in individuals of all races.

The true incidence of RCVS is unknown; clinical experience suggests RCVS is fairly common. [5] Although there is an increasing incidence through reporting due to greater awareness, and use of non-invasive neuro imaging such as computed tomography angiography and magnetic resonance angiography. The treatment of RCVS is still at best observational and no guidelines for treatment has been established for recanalization of the affected vasoconstricted blood vessel.

Case Report

This is a case of a 32 year old, G3P(3003) 2 weeks post-partum, right-handed, married, Filipino, roman Catholic , unemployed , from Valenzuela who was admitted at the Jose R. Reyes Memorial Medical Center on February 3, 2019.

On February 2, 2019 3pm, patient slept asymptomatic. At 9 pm, she was observed by the husband to be moaning in bed with preferential movement of the left extremities and no movement of the right extremities. She was noted to be incoherent and with incomprehensible sounds. There was no fever, no chest pain, no shortness of breath, and no dyspnea. She was immediately brought to the emergency room. Past medical history and family history were unremarkable. She is a non-smoker, non-alcoholic, denies use of illicit drugs.

Assessment

Patient was seen to be incoherent with inappropriate response. Vital signs and general physical examinations were unremarkable. On neuro examination, patient was awake, with global aphasia, homonymous hemianopsia on the right, right central facial palsy, right hemiplegia and Babinski on the right with NIHSS score of 22.

Laboratory workup

Basic laboratory examinations such as CBC, electrolytes and coagulation studies all revealed unremarkable. Other ancillaries like random blood sugar, ESR, CRP and rheumatoid factor, protein C, Protein S and ANA were normal. A cranial MRI with MRA was done showing restricted diffusion on DWI/ADC indicative of acute cerebral infarction and on MRA, there was an area of focal stenosis on the M2 segment on the left middle cerebral artery (Figure 1). Transcranial doppler ultrasound was done revealing decrease mean flow velocities and normal pulsatility indices on the middle to distal segments of the left middle cerebral artery (45-55mm depth) consistent with a focal stenosis (Figure 2). A presumptive diagnosis of reversible cerebral vasoconstriction syndrome was made. She was admitted at the Acute Stroke Unit and given antiplatelet (Cilostazol) and started on physical therapy. Physical rehabilitation was started while admitted in the acute stroke unit. On the 4th day of admission, she had a noted improvement in motor strength to 2/5 and she was able to comprehend and follow simple commands. A repeat transcranial doppler was done revealing improvement to normal values of the mean flow velocities of the middle and distal

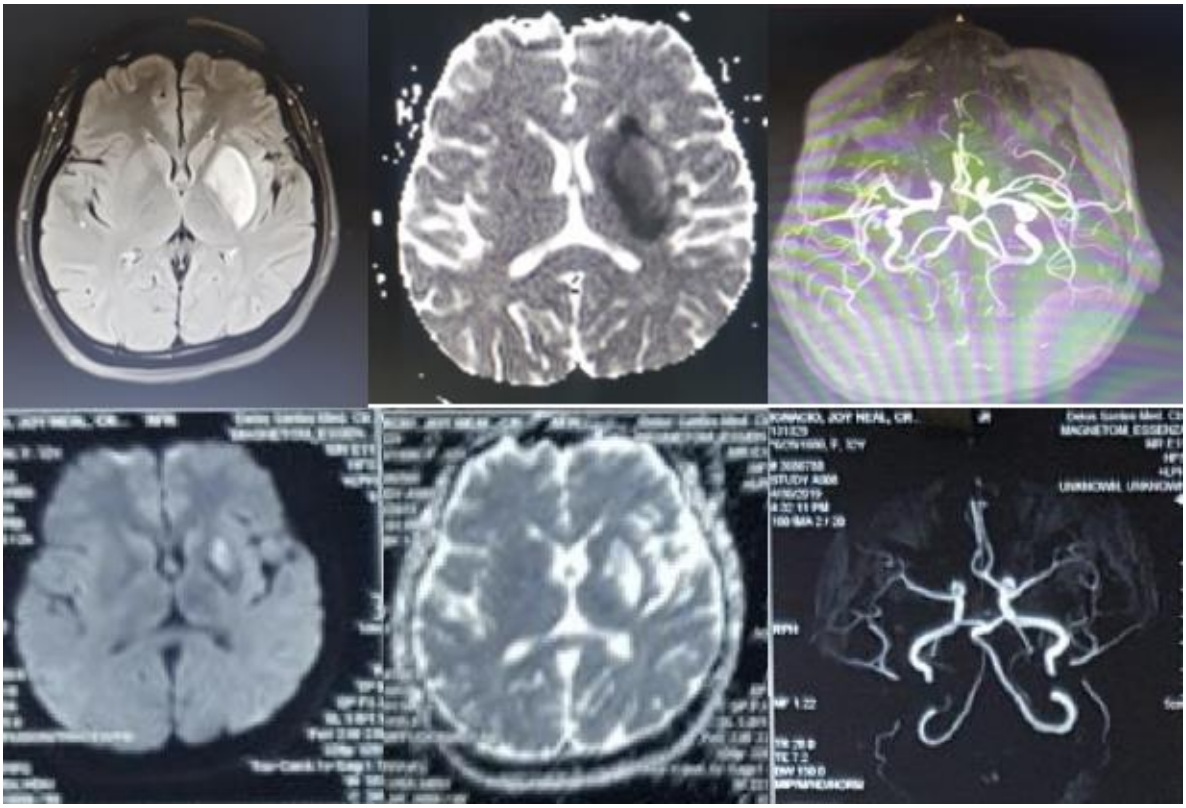

Figure 1 (clockwise from top-left). Upper panel: (1-month post-ictus) Restricted diffusion on the left basal ganglia on DWI. Signal drop on ADC map. Focal stenosis on the middle segment of the left middle cerebral artery. Lower panel: (3 months post-ictus) MRI no focal stenosis on MRA

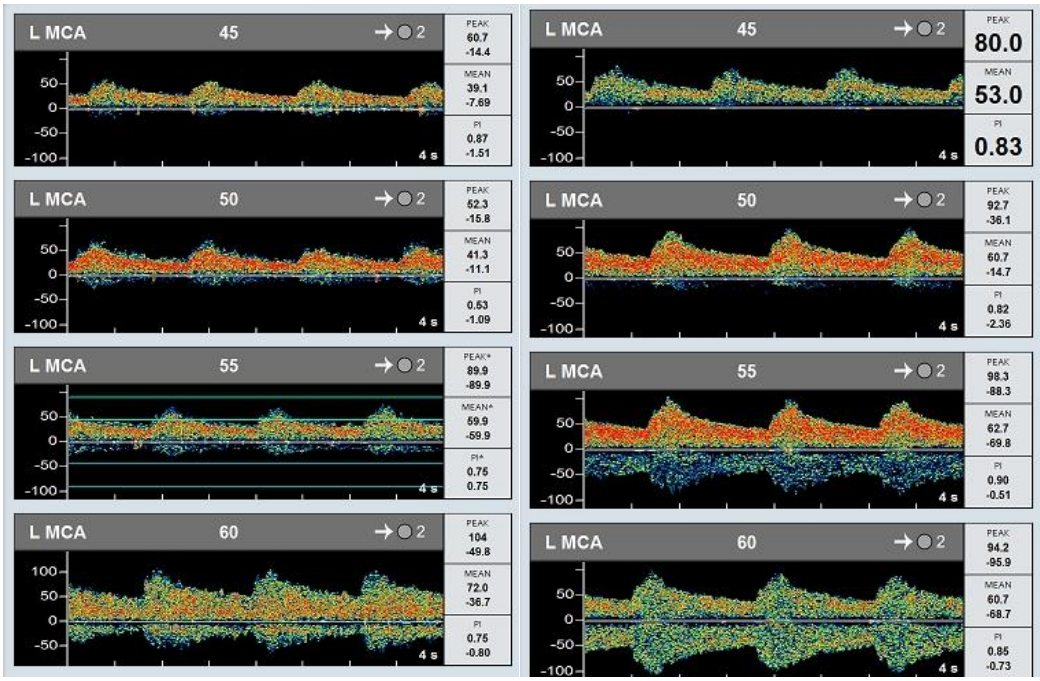

Figure 2. Left: Transcranial doppler sonography (TCD) done 1-day post-ictus showing with dampened waveforms and decreased mean flow velocities in the middle to distal segments of the left middle cerebral artery (45-55mm depth). Right: TCD done 4 days post-ictus revealing improvement of the mean flow velocities of the middle to distal segments of the left middle cerebral artery.

segments of the left middle cerebral artery (Figure 2). The patient was then discharged with an NIHS score of 12 and maintained on cilostazol taken twice a day for 3 months. no adverse drug reactions (bleeding) were reported.

Outcome

On follow up after 2 weeks, there was persistence of right hemiparesis and patient remained wheelchair-bound. Around 14 days post ictus, there was noted improvement in motor strength, she was able to walk with assistance and communicate verbally albeit dysarthric. A repeat cranial MRI with MRA revealed chronic infarct in the left capsuloganglionic region with encephalomalacic changes and mild ex vacuo dilatation on the adjacent left ventricle with no distinct evidence of stenosis, aneurysm or vascular malformation. (Figure 2) On the 90 days post-ictus, she followed-up able to walk with cane, communicates fluently with an MRS score of 3.

Discussion

RCVS has been associated with a variety of conditions including pregnancy [6], migraine [7] use of vasoconstrictive drugs[8] and neuro-surgical procedures[9], hypercalcemia[10], and unruptured saccular aneurysms [11].

Some authors have speculated that the vasoconstriction is related to transient vasculitis, but there is no evidence to support a role for inflammation. Cerebrospinal fluid examination and extensive serological tests are normal, and pathological studies of the brain and temporal arteries have shown no abnormality[12].

The most common clinical presentation of RCVS is headache which was not present in the case presented. This is usually thunderclap in character and lasts for seconds to days with intermittent recurrence. Owing to the incidence of RVCS as subarachnoid hemorrhage. This case has an atypical and exceptional presentation due to no presence of headache. Investigation for RCVS would involve triggering factors such as orgasm, physical exertion, acute stress and Valsalva maneuvers, bathing and swimming which is also not present in the case. The symptom in this case is a development of a focal neurologic deficit, presented as weakness on the right upper and lower extremities, right hemianopsia and global aphasia with a documented acute left basal ganglia infarction on neuroimaging[13].

The most common presentation of RCVS as stroke is still headache. Typical headache is bilateral (although it can be unilateral), with posterior onset followed by diff use pain[18]. Lesions are noted in 12–81% of patients, dependent on patterns of study recruitment. Typically RCVS will present as subarachnoid hemorrhage on non-enhanced cranial CT scan which shows cortical sulcal space surface hyperdensity[14, 15]. These are mostly present among individuals with RCVS below 60 years old. [16]

In the review made by Singhal (2019) infarctions attributable to RCVS were observed to be bilateral, symmetrical, and in borderzone areas. In the same study, infarctions due to RCVS may also present as larger occlusions in the cerebral arteries present wedged shaped infarctions.

Since there was an uncertainty about the cause of cerebral arteriopathy, there is a need for obtaining complete blood count, electrolytes, liver and renal function tests, and tests for inflammation (e.g., erythrocyte sedimentation rate, rheumatoid factor, and antinuclear cytoplasmic antibodies), all of which are typically normal in patients with RCVS.

The International Headache Society provided a criteria for acute reversible cerebral angiopathy. This includes 1. Acute and severe headache (often thunderclap) with or without focal deficits or seizures 2. Uniphasic course without new symptoms more than 1 month after clinical onset 3. Segmental vasoconstriction of cerebral arteries shown by indirect (e.g., magnetic resonance or CT) or direct catheter angiography 4. No evidence of aneurysmal subarachnoid hemorrhage 5. Normal or near-normal CSF CSF (protein concentrations < 100 mg/dL < 15 white

blood cells in uL 6. Complete or substantial normalization of arteries shown by follow-up indirect or direct angiography within 12 weeks of clinical onset. Of which, only the segmental vasoconstriction, no aneurysmal subarachnoid hemorrhage and complete resolution of arteries by 90 days were seen in this case. In the study of Singhal (2007) A diagnosis of RCVS can only be confirmed when the reversibility of the vasoconstriction is assessed; 12 weeks from onset of symptoms has been proposed as a cutoff by which reversal should be complete or at least substantial. In the case presented, uniphasic course, normal laboratories, segmental vasoconstriction and complete normalization of arteries was seen in this case.

The pharmacologic treatment for RVCS on observational studies include calcium channel blockers and brief courses of magnesium sulfate. The use of nimodipine in some studies does not necessarily affect the time course of cerebral vasoconstriction but has been seen to relieve the intensity and recurrence of headache. The vasodilatory effect of phosphodiesterase inhibitor 3 (Cilostazol) was applied for this case to address the focal stenosis documented on angiography. for 3 months was used in this case to address the focal stenosis seen on ang and was seen to have improvement of motor power with concomitant rehabilitation. There was recanalization of the affected vasoconstricted area and a significant reduction of NIHSS and Modified Rankin scale. [17]

Conclusion:

We report a rare case of a young female 2 weeks post-partum, with no co-morbidities presented with focal neurologic deficit, with acute infarction and stenosis on angiography and remarkable improvement both clinically and radiologically (angiography). Although infarction is a rare presentation, an impression of RCVS should be considered as a diagnosis.

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Ethical Consideration

Patient form was secured before submission of manuscript

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