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A Rare Case of Reversible Cerebral Vasoconstriction Syndrome in a Patient with Migraine

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Abstract

Reversible Cerebral Vasoconstriction Syndromes (RCVS) are a cluster of conditions that show reversible multifocal vasoconstriction of the cerebral arteries with typical manifestations like thunderclap headache and less commonly focal neurologic deficits related to brain edema, stroke, or seizure.

We describe a case of 31 year old male staff nurse who arrived at the emergency room for regular morning duty and suddenly became unresponsive. He was immediately intubated and managed following which NCCT brain was taken. It revealed generalized brain edema. Other blood and radiological investigation reports were unremarkable. After ruling out all possible diagnosis he was diagnozed as a case of RCVS. He was well-managed by the emergency team and was discharged in a stable condition with nil neurological deficit.

Keywords: RCVS; Thunderclap headache; Migraine; Stress; Sausage on a string

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Introduction

Reversible Cerebral Vasoconstriction Syndrome (RCVS) typically presents with excruciating headaches, with or without other concomitant neurological signs and symptoms, and diffuse segmental constriction of cerebral arteries that resolve spontaneously within 3 months.[1,2] Thunderclap headache is usually the initial symptom and commonly reoccurs for 1–2 weeks.[3,4] Ischemic and hemorrhagic stroke are the predominant sequelae of this syndrome.[4] Hereby we are reporting a case of Reversible Cerebral Vasoconstriction Syndrome (RCVS) triggered by vasoactive drug and stress.

Case Study

31 year old male staff nurse arrived at the emergency room for usual morning duty. After some time, he was found unresponsive in the resuscitation area.

Primary assessment showed,

Airway– unstable

Breathing– RR-5bpm, SPO_2-30%, No abnormal sound

Circulation- PR 138bpm, regular, blood pressure -200/110 mmhg, Peripheries cyanosed.

Disability- GCS: E1 V1 M1, Pupil dilated bilateral and not reactive, no nystagmus.

Patient airway was immediately secured with endotracheal intubation without giving any sedative and paralytic agent. Labetalol 20mg IV stat was then given. After that patient's SpO_2 improved to 100% almost immediately following which patient was shifted to NCCT brain to rule out intracranial hemorrhage. NCCT brain showed generalized brain edema (Figure 1). Other than that no significant abnormality was noted.

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All the basic blood investigations, cardiac enzyme, Urine vanillylmandelic acid, blood and urine tox screening were sent. Later they were deduced to be within normal limit. MRI angiogram brain showed essentially vessels of neck and brain with no remarkable pathology and no obvious aneurysm/AV malformation or any flow limiting stenosis.

Until then patient wasn't responsive. After that patient started moving his leg and hence was sedated with midazolam. MRI Venogram was essentially a negative study with unremarkable venous sinuses of brain. ECG showed sinus tachycardia. 2D ECHO showed no RWMA and LVEF at 55%.

Patient was consequently managed with anti hypertensive and IV fluids and admitted under Dept. of Neurology in ICU. Transcranial Doppler was also normal. EEG too remained unremarkable. By afternoon the same day patient's sedation was discontinued and patient was extubated. CSF analysis was also done to rule out any infection or SAH after 12 hours. It showed nil deviations from normality. After that from the fully conscious patient, a retrospective history was taken.

He revealed that he is Suffering from of migraine since 10 years and that he used to get migraine attacks usually once or twice every month. Whenever he had severe headaches he was consuming Naxdome 500mg [Naproxen + Domperidone] and Vasograin [Ergotamine (1 mg) + Caffeine (100 mg) + Paracetamol (250 mg) + Prochlorperazine (2.5 mg)]. But in the past 2 weeks the episodes were increased as much as 2 times a week and also the headaches got intensified.

A history of considerable work stress was also there due to hectic emergency duties the past two weeks. So, the day before the incident he had severe headache and asusual he took Vasograin and Naxdome. But the pain failed to settle down with the medications. So, he loaded him with further parenteral Tramadol in a nearby local hospital.

Subsequently at the day of incident he came to the hospital quite normal. After that he felt severe headache, blurring of vision, profuse sweating, choking and inability to move all his limbs following which he became unresponsive.

Two days later patient was discharged in stable condition with nil neurological deficit.

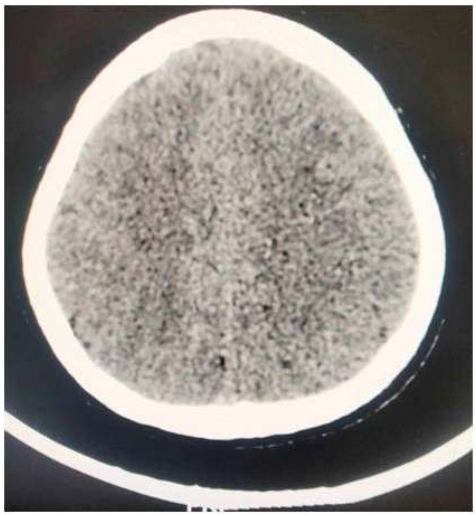

Fig. 1: NCCT brain shows generalized brain edema

Discussion and Therapeutic Considerations

The actual prevalence of RCVS is unknown. The disease predominantly affects individuals between 20 and 50 years of age.[5] However, children can be affected as well. RCVS occurs more commonly in women than men.[6]

The precise etiopathogenesis of RCVS continuous to be uncertain to date; however, sympathetic hyperactivity might be involved.

RCVS can occur spontaneously or precipitated by a myriad of factors. Vasoactive drugs [e.g.,: cannabis, cocaine, nasal decongestants, Triptans, Ergot alkaloid[7] derivatives, SSRI and the postpartum time are frequent associations.[8] RCVS has can also been chained to the intake of immunosuppressant and cytotoxic drugs and Catecholamine-secreting tumours.

Many patients report trigger factors, such as physical exhaustion, orgasm, sudden emotional disturbances straining, coughin, bathing, and swimming.

Clinical features

Headache is the chief complaint and often remains the defacto presentation of RCVS.[6] The headache tends to be recurrent in up to 94–100% of patients. Emesis, nausea, disorientation, blurring of vision and transiently elevated blood pressure frequently adjoins the headache. Focal neurological deficits and generalised tonic–clonic seizures can manifest too.[8]

The following criteria have 98 to 100 percent specificity and a similarly high positive predictive value andhence can be exploited for rapid bedside diagnosis bypassing the need of cerebral angiography or documentation of vasoconstriction reversal on follow-up imaging:

- Recurrent thunderclap headache, (or)
- Single thunderclap headache in combination with either normal neuroimaging, border zone infarcts, or vasogenicedema, (or)
- No thunderclap headache but abnormal angiography and no brain lesions on neuroimaging.

Laboratory findings

Routine tests→ Hemogram, serum electrolytes, liver and kidney function test

Tests for inflammation→ ESR, rheumatoid factor, and antinuclear cytoplasmic antibodies are usually normal in RCVS.

Urine vanillylmandelic acid and 5-hydroxyindoleacetic acid → To rule out pheochromocytoma, carcinoid

Blood and urine tox screens→ To rule out illicit drug use

CSF→ normal (ie, protein level <60 mg/dL, ≤5 white blood cells per mm^3) in more than 85 percent of patients [8]; minor abnormalities may be seen due to hemorrhagic stroke.

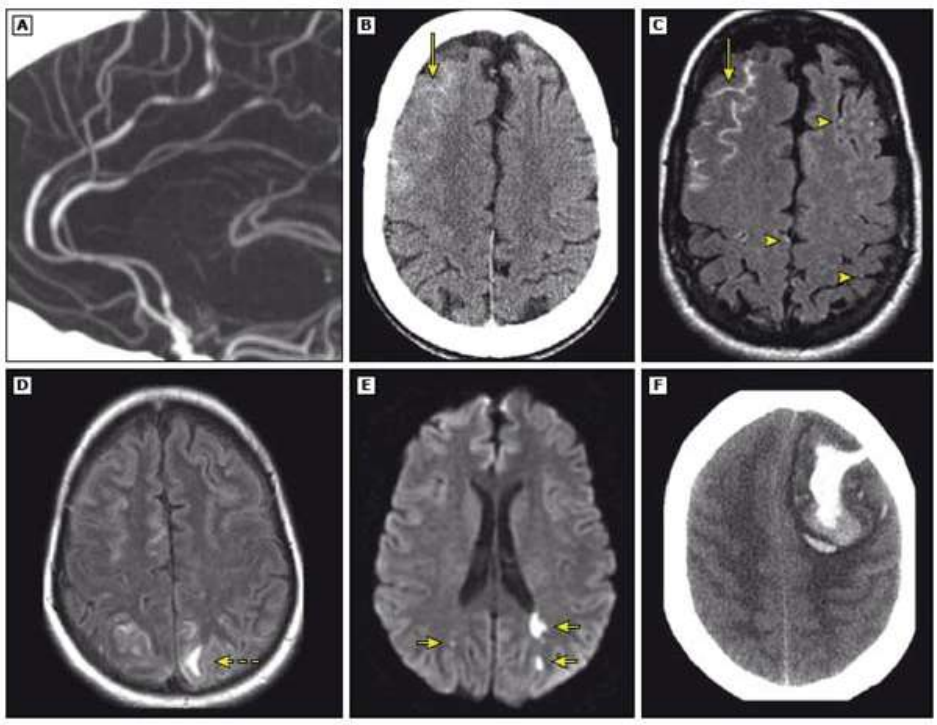

Fig. 2: (A) Head CT angiogram: sagittal image showing the classic "sausage on a string" appearance of both anterior cerebral arteries.
(B) Head CT: axial image showing subarachnoid hemorrhage (SAH) overlying the right frontal lobe (arrow).
(C) Brain MRI: axial FLAIR image showing the right frontal SAH (arrow) as well as multiple dot-shaped hyperintensities (arrowhead) within the sulci of both hemispheres, suggesting the presence of dilated cortical surface arteries.
(D) Brain MRI: axial FLAIR image showing the posterior-predominant crescentic hyperintense signal in the cortical-subcortical regions (dashed arrow). Corresponding DWI and SWI (not shown) were normal. These findings suggest the presence of brain edema as described in the posterior reversible leukoencephalopathy syndrome.
(E) Brain MRI: axial DWI showing ischemic lesions (short arrows) in the bilateral "watershed" regions of the middle and posterior cerebral arteries.
(F) Head CT scan: axial image showing a left frontal parenchymal hemorrhage.[8]
RCVS: Reversible cerebral vasoconstriction syndrome; FLAIR: Fluid-attenuated inversion recovery; DWI: Diffusion-weighted image; SWI: Susceptibility-weighted image.

Imaging

Inspite of the generalized cerebral vasoconstriction, initial brain scan is normal in over 50 percent of patients at the time of admission with RCVS. In the subsequent days, many patients progress complications notably,

- ischemic strokes,
- convexal (nonaneurysmal) subarachnoid hemorrhages,
- lobar hemorrhages, and
- Reversible brain edema, solely or in combination.[4,8]
- Cerebral angiographic abnormal features are dynamic and spread proximally, causing a "sausage on a string" appearance of the Circle of Willis arteries and their branches. These abnormalities resolve spontaneously over a few weeks.

Transfemoral, CT, or MR angiography (MRA) can be used to ascertain the segmental cerebral arterial narrowing and vasodilatation.

Treatment

There is no validated treatment for RCVS. Supportive treatment is projected on controling blood pressure, excruciating headaches, and other complications such as seizures. Per oral calcium channel blockers are generally given to revert vasoconstriction but the substantiating evidence for this treatment is not upto the mark.

Triptans and the ergot derivatives are contraindicated because of their vasoconstrictive actions.[9,10] Glucocorticoids may be associated with worse outcome in RCVS.[8,11]

The clinical outcome is mostly benign. Rarely patients develop severe irreversible deficits or death from progressive strokes or cerebral edema. Recurrence of an episode of RCVS is rare.

Migraine

People with prediagnosed RCVS often have notable migraine episodes in the past and this predisposes to the danger of confusing a RCVS episode to be yet another but severe migraine attack.[12,13] Headaches in RCVS are secondary (ie, symptomatic), whereas migraine is a primary headache.[1] People living with migraine who developed RCVS could differentiate the thunderclap headaches from the usual migraine episodes.[4] Migraine appears to be a predisposing cause of hemorrhage during RCVS.[14] Migraine attack treatments such as triptans and ergots can instigate RCVS or increase the vasoconstriction when used to subside a thunderclap headache.[9,15]

Conclusion

RCVS is an uncommon disease and is difficult to diagnose; but because it can cause fatal complications, such as cerebral hemorrhage or cerebral infarction, a robust and continued monitoring is essential. RCVS should be kept in mind in any patient who presents with repeated thunderclap headaches or idiopathic stroke, most notably postpartum or following the intake of vasoactive drugs. The vasoconstrictive drugs used in general migraine aggravate RCVS even further, and so it is crucial to differentiate it from symptoms of migraine.

In our case of RCVS, Vasoactive drugs like ergotamine and stress were the suspected triggers and the likely cause of the condition. Prompt evaluation and early recognition of this condition can help in the good management of RCVS patients. Awareness regarding RCVS and it's triggering factor may further help increase early detection of future RCVS cases.

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