An Unusual case of Reversible Cerebral Vasoconstriction Syndrome

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Abstract:
A 25 year old pregnant lady (36 weeks of amenorrhea) presented with acute onset headache, vomiting followed by generalised seizures and progressive altered sensorium over 24 hours. Her Blood pressure was 190 110mmHg and was diagnosed to have eclampsia, initiated on Magnesium sulfate and underwent lower segment caesarian section. She was imaged with MRI-MRA with diffusion study which showed right occipital and bilateral cerebellar hyperintensities with areas of restriction of diffusion and diffuse narrowing of vessels of the vertebrobasilar system. There was distortion of brainstem and 4th ventricle with obstructive hydrocephalus. She underwent an External Ventricular Drainage (EVD) followed by posterior fossa decompressive craniectomy. Over 96 hours, her sensorium had improved to a GCS of 15. Repeat MR Imaging showed resolution of cerebellar hyperintensities and normalization of the diffusion restriction, with normal vessels on MR angiogram. Subsequent transcranial doppler of vertebrobasilar arteries was also normal. Based on the clinical setting of headache, raised blood pressure in the peripartum period and the serial imaging findings, reversible cerebral vasoconstriction syndrome was diagnosed. The atypical features in this case were the hyper acute presentation progressing to coma within 24 hours, the initial MRI showing diffusion restriction, with significant mass effect and obstructive hydrocephalus requiring posterior fossa decompression and disease process being confined to the posterior circulation.

Keyword:
Reversible Cerebral Vasoconstriction Syndrome (RCVS), Reversible posterior leukoencephalopathy (RPLE), Thunderclap headache

INTRODUCTION
Reversible Cerebral vasoconstriction syndrome (RCVS) consists of sudden, severe headaches with/without associated
neurologic deficits occurring as a result of reversible multifocal narrowing of the cerebral arteries.[1][2] RCVS is usually self-limiting with resolution of headache and vasocostriction occurring over days to weeks. [2] Diagnostic criteria include severe, acute headaches, with or without additional neurologic signs or symptoms, normal cerebrospinal fluid analysis, exclusion of other causes of sudden severe headache, multifocal segmental cerebral arterial vasocostriction on angiography, and reversibility of the vasocostriction within 12 weeks after onset.[3] In this article, we report an unusual presentation of Reversible cerebral vasocostriction syndrome.

CASE REPORT:
A 25 year old pregnant lady (36 weeks of amenorrhea) presented with acute onset of severe holocranial headache with projectile vomiting, followed by seizures and progressive altered sensorium over the next 24 hours. Her past medical history was unremarkable. Her antenatal period was uneventful without hypertension or albuminuria. On examination, she had GCS - 6/15, blood pressure of 190/110 mm of Hg, 3 mm pupils with sluggish light reaction and papilloedema without focal deficits. Meningeal signs were absent. She was taken up for lower segment caesarean section, with a provisional diagnosis of Eclampsia. Other differentials considered were cerebral venous thrombosis, cerebrovascular accident and reversible posterior leukoencephalopathy (RPLE). In the immediate postoperative period, her GCS was 2T/15 with dilated and nonreactive pupils. CT Brain showed hypodensity in the cerebellar hemispheres with mass effect on the brain stem causing obstructive hydrocephalus (Figure 1A). She underwent an emergency right side EVD procedure with which her GCS improved to 5T/15 (E2M3VT). Subsequently, a posterior fossa decompressive craniectomy was done. MRI Brain showed T2 hyperintensities in both the cerebellar hemispheres and occipital lobes with restricted diffusion. The cerebellum was swollen with mass effect on the brainstem. MRA showed narrowing of the vessels of the vertebrobasilar system (Figure 1B-F). Her sensorium improved gradually to 15/15 over 96 hours. Three weeks after surgery, she was ambulant independently with minimal limb incoordination. Her neck and transcranial doppler study done during the resolution phase (three weeks into illness) were normal. Repeat MRI brain done three months after the acute event showed resolution of hydrocephalus, cerebellar hyperintensities and diffusion restriction with a normal MR angiogram (Figure 2A-E). In the setting of peripartum period, considering the acute onset severe headache, raised blood pressure, MRI brain/ MRA changes and their resolution in subsequent Doppler and MRI with MRA, RCVS was diagnosed.

DISCUSSION:
RCVS is a unifying term for a group of diverse conditions (Call Fleming syndrome, Thunderclap headache with reversible vasospasm, benign angiopathy of the CNS, postpartum angiopathy, Migrainous vasospasm/ angiitis, Drug-induced cerebral angiopathy).[4][5] RCVS is due to transient dysregulation of cerebral vascular tone leading to segmental/ multifocal arterial constriction/ dilatation, starting in distal arteries and then progressing towards moderate to large arteries.[2] Neurovascular imaging in RCVS shows multifocal, segmental vasocostriction with areas of dilatation involving large/ medium sized
vessels of both anterior and posterior circulation. RCVS can occur spontaneously, or secondary to exposure to vasoactive substances, and/or related to postpartum state. RCVS is seen in 20-50 years age group, with female preponderance. It usually presents as severe acute headache, often thunderclap headache, with or without emesis, photosensitivity, focal deficits and seizures. Headaches tend to recur over days to weeks, with other neurological symptoms including transient visual/sensory symptoms, aphasia, hemiparesis, and ataxia developing later. Impairment of consciousness is infrequent and usually mild, but coma may occur in rare cases with multiple strokes. Transient hypertension may be seen. Our patient had acute onset of headache, followed by seizures, progressive altered sensorium and then became comatose, which is not like the typical recurrent thunderclap headaches followed by deficits over few days as seen in classical RCVS. Brain parenchymal imaging at baseline can be normal or can show cortical subarachnoid hemorrhage (cSAH) or intracerebral hemorrhages (ICH). Ischemic strokes particularly in watershed regions occur typically in the second week. Our patient had showed T2 hyperintense lesions in both the Cerebellar hemispheres and occipital lobes with scattered punctuate areas of restricted diffusion in cerebellum. An unusual feature in this case was the cerebellar swelling causing mass effect on the brainstem, obstructive hydrocephalus, and progressive worsening in sensorium requiring posterior fossa decompressive craniectomy, which has not been described in literature. Another rare finding was the MRA showing irregular narrowing of the major arteries of the vertebrobasilar system and their branches, with minimal changes in the arteries of the anterior circulation. RCVS is usually a benign, self-limiting condition with resolution of headaches and complications include transient ischemic attacks (up to 16%), RPLE (9-14%), ischemic strokes (4-54%), cSAH (up to 34%), ICH (up to 20%) or subdural hemorrhage (2%) and seizures (up to 21%). Focal neurological deficits are found in 9-63%, permanent deficits in 9-29% and mortality in a few patients. Hemorrhage or infarction are associated with poor outcome. In our patient, repeat brain imaging after 12 weeks showed decrease in the T2 signals, normalization of the earlier diffusion imaging changes and resolution of hydrocephalus/mass effect and a normal MR Angiogram. Neck and transcranial Doppler study was also normal, suggesting resolution of the vascular changes. In our case, the clinical course was of a hyper acute presentation progressing to coma within 24 hours, initial MRI showed diffusion restriction, mainly in the cerebellum, significant mass effect and hydrocephalus requiring posterior fossa decompression, and vascular changes being restricted to the posterior circulation vessels. These features were in contrary to the typical features of RCVS described in literature.

**CONCLUSION**

This case illustrates that RCVS can have a hyper acute presentation rapidly progressing to coma within 24 hours. The pathology may be restricted only to the posterior circulation. Significant mass effect due to oedema and resultant hydrocephalus warrant timely surgical interventions like posterior fossa decompression. Even with a poor clinical status and significant MRI changes, patients with RCVS can have a potentially good outcome. Posterior fossa decompressive craniectomy done at right time has prevented the...
mortality in this case.

REFERENCES


Figure legends

Figure 1: CT Brain (1A) showed bilateral Cerebellar hypo densities (arrow) and hydrocephalus (arrow heads). MRI Brain showed T2 hyperintensities in Cerebellum (1B, arrow) and occipital lobes (1C, arrow) with restricted diffusion on DWI-ADC images (1D-E, arrows). MRA showed narrowing of both vertebral/ basilar arteries (1F, arrows).

Figure 2: MR T2 Axial Brain showing resolution of the previously seen hyperintensities in cerebellum (Figure 2A, arrow) and occipital lobes (Figure 2B, arrow), and normalisation of the DWI-ADC (Figure 2C-D, arrow) and MRA changes Figure 2E).