CASE REPORT

Reversible cerebral vasoconstriction syndrome and bilateral vertebral artery dissection presenting in a patient after cesarean section

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SUMMARY

Reversible cerebral vasoconstriction syndrome (RCVS) is characterized by sudden-onset thunderclap headache and focal neurologic deficits. Once thought to be a rare syndrome, more advanced non-invasive imaging has led to an increase in RCVS diagnosis. Unilateral vertebral artery dissection has been described in fewer than 40% of cases of RCVS. Bilateral vertebral artery dissection has rarely been reported. We describe the case of a patient with RCVS and bilateral vertebral artery dissection presenting with an intramedullary infarct treated successfully with medical management and careful close follow-up. This rare coexistence should be recognized as the treatment differs.

BACKGROUND

Reversible cerebral vasoconstriction syndrome (RCVS) is a syndrome characterized by sudden-onset thunderclap headache and focal neurologic deficits, most commonly in women aged 20–50 years.^{1–2} Different names are given to this syndrome according to clinical context including Call–Fleming syndrome, benign angiopathy of the CNS, postpartum angiopathy, thunderclap headache with reversible vasospasm, migrainous vasospasm or angiitis and drug-induced cerebral arteritis or angiopathy.² In general, these disorders have been poorly characterized and continue to be frequently confused with cerebral vasculitis due to overlapping angiographic features and, to a certain degree, clinical features.³ Common imaging findings are diffuse segmental cerebral vasoconstriction of the intracranial internal carotid arteries, basilar artery and arteries of the circle of Willis that spontaneously resolves in weeks to months.

Although the pathophysiology of RCVS remains unknown, the prevailing hypothesis involves a transient disturbance in vascular tone.^{2–4} This alteration in vascular tone may be spontaneous or evoked by various exogenous or endogenous factors. Pregnancy-related hormonal effects are known to alter vascular tone.⁵ Although reports of RCVS are found in the literature, only a few authors have reported on RCVS and an association with vertebral artery dissection.⁶

We report a case of bilateral vertebral artery dissection-associated RCVS in which the patient presented 10 days post partum.

CASE PRESENTATION

A previously healthy middle-aged woman presented to an outside facility 10 days post partum with shortness of breath, chest discomfort and headache. A CT of the head was performed and interpreted as normal. A magnetic resonance venogram (MRV) was performed and showed no intracranial venous sinus thrombosis. Following the MRV the patient started to complain of facial paresthesias, right-sided dysmetria and worsening headache. An MRI was performed which revealed an intramedullary infarct with no supratentorial foci of abnormal T2 or diffusion signal. A CT angiogram showed an abnormal appearance of the right vertebral artery without associated subarachnoid hemorrhage or pseudoaneurysm. Cerebrospinal fluid (CSF) studies were negative for findings of inflammation or infection. At this time the patient was referred to our facility.

At presentation to our facility she complained of dizziness, headache localized to the posterior neck and occasional stabbing posterior right calvarial pain. Past medical history was remarkable for recent childbirth via cesarean section and migraines since the age of 7 years that were well-controlled without need for prophylactic medication. This headache was different from her normal migraine which consisted of throbbing unilateral pain, nausea/vomiting and photophobia. Laboratory tests and social history were non-contributory. Physical examination was remarkable for horizontal left beating nystagmus when looking up or to the right, decreased right eye papillary constriction, right eye ptosis, right finger to nose dysmetria, decreased right hemiface sensation and left extremity thermoanesthesia. Blood pressure was normal.

INVESTIGATIONS

Cross-sectional imaging demonstrated no abnormal signal intensity in the supratentorial brain (figure 1A). Evaluation of the infratentorial brain confirmed the presence of an intramedullary infarct, now subacute (figure 1B, C). The vasculature was remarkable for the suggestion of mild posterior circulation irregularities (figure 2C) and bilateral vertebral artery dissection (figure 3A, D). Given the rare occurrence of bilateral vertebral artery dissection in conjunction with posterior circulation vasospasm, catheter angiography was performed to assess for other potential etiologies such as pseudoaneurysms or dissection aneurysm which may not be as readily detected on CT angiography. Cerebral angiography confirmed bilateral dissection and mild proximal posterior circulation vasospasm

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Figure 1 Intramedullary Infarct. (A) MR images of the brain obtained on a 1.5 Tesla magnet demonstrating a normal appearance of the white matter and basal ganglia on FLAIR imaging. (B) Axial T2 fast spin echo images through the level of the medullary cord demonstrating a right lateral margin focus of increased T2 signal suggesting tissue injury. (C) Consistent with injury several days previously, there was no restricted diffusion signal in this lesion. The overall imaging features were consistent with a subacute infarct.

(figures 2A, B and 3B, E). Transcranial Doppler evaluation was performed on the same day to establish a baseline velocity for comparison and to allow non-invasive monitoring of perfusion at the bedside. It showed that the anterior cerebral circulation velocities were within normal limits and the posterior cerebral circulation velocities were mildly increased.

DIFFERENTIAL DIAGNOSIS

The differential diagnoses for focal intracranial vessel irregularities may include atherosclerotic disease, vasculitis (either inflammatory or infection) and vasospasm (either idiopathic or secondary to hemorrhage). The differential was extremely narrowed by the patient's normal CSF results which excluded hemorrhage and vasculitis either due to inflammation or infection. Given the patient was young, in otherwise good health and with a relatively normal lipid panel, atherosclerotic disease was excluded. The proximal nature of the spasm favored that of RCVS over autoimmune vasculitis, which is often more distal in distribution. Migrainous spasm was also considered less likely due to the persistent nature of the patient's symptoms as well as the associated stroke.

TREATMENT

Given the bilateral nature of the patient's disease and the lack of significant flow degradation, it was felt that endovascular treatment of her spasm/dissection may do more harm than good. Edwards *et al*⁷ demonstrated a low stroke rate in patients with blunt cerebral vascular injury on dual-therapy treatments, so medical management with aspirin and clopidogrel was initiated while the patient was in the high risk for dissection progression window. After 1 week she was discharged home with improved dysmetria but persistent right ptosis/miosis and decreased right hemiface sensation. Short interval clinical evaluation demonstrated stable clinical findings and the patient was transitioned to monotherapy with aspirin alone.

OUTCOME AND FOLLOW-UP

At 6-month follow-up her symptoms had improved overall with a slight residual right ptosis, mild decreased hemifacial sensation and mild left extremity thermoanesthesia. It was decided to use CT angiography rather than MR angiography for re-evaluation due to the higher spatial resolution and improved sensitivity/specificity when assessing the vertebral arteries.⁸ The follow-up CT

angiogram demonstrated no residual dissection or spasm (figures 2D and 3C, F).

DISCUSSION

RCVS is a complex entity without a well-understood pathophysiology. Pregnancy, migraines, exposure to certain vasoactive drugs and trauma are medical conditions that have been associated with RCVS. The clinical course of RCVS is usually benign without treatment; however, in some cases severe neurologic dysfunction can occur due to ischemia in regions perfused by severely contracted arteries, cortical subarachnoid hemorrhage, intracerebral hemorrhages and/or reversible posterior leukoencephalopathy.¹

This case was unique for several reasons. Most patients who present with RCVS complain of severe onset of headache. Although our patient presented with headache, it was not of sudden or severe onset and was confounded by the history of migraines leading to a longer time period between presentation and cross-sectional imaging. The history of migraines and non-specific angiographic findings of RCVS also complicated the diagnosis. Multisegmental vasoconstriction has been reported with migraines in the postpartum period, following sexual intercourse, following aneurysmal subarachnoid hemorrhage and after administration of drugs.¹ Similar angiographic findings can also be seen with cerebral vasculitis or primary CNS angiitis; however, they classically present with gradually progressive neurologic symptoms, encephalopathy and CSF abnormalities in 80–90% of cases.¹ Continued work in MRI is making headway in trying to separate RCVS from other etiologies. Increased signal in fluid-attenuated inversion recovery sequences has been seen in cases of RCVS.⁹ Contrast-enhanced high resolution axial MR images have been used with some success in differentiating RCVS from vasculitis.¹⁰ Unfortunately, neither was definitive in our case. Our patient's negative CSF studies, mildly increased transcranial Doppler velocities and resolution of angiographic findings were consistent with reversible vasospasm.

Despite no clear mechanism, an association between spontaneous carotid/vertebral artery dissection and RCVS in the postpartum period has been suggested.⁶ Hormonal effects on the vessel wall including smooth muscle cell proliferation, decreased collagen synthesis, eosinophilic infiltration and alterations in cell wall formation have been suggested as etiologies for dissection.⁵

Figure 2 Cerebral vasospasm. Diagnostic angiogram images in the arterial phase of (A) the right and (B) the left vertebral artery showing narrowing of the distal right V4 segment (black arrow) and proximal left P1 segment (black arrowhead) as well as asymmetric filling of the posterior circulation. (C) Volume rendered images from a CT angiogram performed the same day show similar findings (white arrow and white arrowhead). No definite pseudoaneurysm was identified nor were there findings of intracranial subarachnoid hemorrhage. (D) Six-month follow-up CT angiogram shows resolution of the distal right V4 segment focal narrowing and proximal left P1 segment.

Increased cardiac output and increased blood volume may contribute to increased shear stresses and subsequent dissection.⁵ Spontaneous dissections have also been seen in patients after epidural anesthesia.^{5–11} Cases that presented in the peripartum period either presented immediately¹² or 5–18 days after routine vaginal delivery.⁶ One factor that seems to hold true for most cases in the literature is the history of migraines. Fortunately, a distinct difference between the normal migraine headache and the headache at the time of dissection was reported.⁶

Our patient's mild non-flow-limiting vasospasm and bilateral dissections, which may suggest increased vessel friability, led to the decision to treat conservatively with careful clinical follow-up and observation. Vertebral dissections are often treated with conservative medical management while the optimal management of RCVS may include angioplasty, calcium channel blockers, glucocorticoids, magnesium sulfate and/or

observation.¹ Unfortunately the clinical course may be devastating; symptoms may clear, only to return within days.¹ The return of symptoms may include infarction, most commonly in watershed areas.¹ Fortunately, conservative treatment did not lead to a worsening of symptoms in our patient. If this patient had shown a progressive decline in her examination or change in transcranial Doppler/perfusion studies, we would have proceeded back to the catheter laboratory for consideration of intra-arterial verapamil or nicardipine and/or angioplasty.

In conclusion, this is an example of a patient who presented with symptoms of an intramedullary stroke secondary to RCVS and bilateral vertebral artery dissection and was treated successfully with conservative management. Radiologists should be aware of the association between pregnancy, RCVS and dissection and should have a low threshold to use MRI to evaluate for findings of RCVS and dissection as the treatment of RCVS versus dissection may be different.

Figure 3 Vertebral artery dissection. Curved planar reconstruction CT image with contrast demonstrating irregularity and narrowing of (A) the right and (D) the left vertebral artery throughout their cervical course. Diagnostic cerebral angiogram performed within 24 h of the CT angiogram confirmed vertebral artery dissection bilaterally (right, B; left, E) and excluded pseudoaneurysm or dissecting fusiform aneurysm. Six-month follow-up CT angiogram curved planar images show resolution of the vertebral artery irregularities (right, C; left, F).

Learning points

- There is an association suggested between pregnancy, RCVS and dissection.
- Most cases of RCVS-associated dissection involve the carotid vessels.
- Differentiating between RCVS and dissection is important as treatment options differ between the two pathologies.
- RCVS may be treated conservatively with medical management and close imaging/clinical follow-up.

Competing interests None.

Patient consent Obtained.

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