CASO CLÍNICO/CASE REPORT

Superior Cerebellar Artery Dissection in a Patient Diagnosed with Reversible Cerebral Vasoconstriction Syndrome: A Case Report

Disseção da Artéria Cerebelosa Superior numa Doente Diagnosticada com Síndrome de Vasoconstrição Cerebral Reversível: Um Caso Clínico

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Abstract

Arterial dissection is an uncommon complication of reversible cerebral vasoconstriction syndrome (RCVS). We describe the case of a 35-year-old woman with a migraine history who presented with recurrent thunderclap headache and focal neurological signs, including right hemiataxia. She had been diagnosed with COVID-19 disease two weeks earlier. Neuroimaging revealed multifocal stenosis of the posterior circulation arteries and dissection of the right superior cerebellar artery. She improved significantly throughout her one-week hospitalization and maintained only mild ataxia. The interplay between COVID-19 disease, RCVS, and arterial dissection requires further investigation.

Resumo

A dissecção arterial é uma complicação rara da síndrome de vasoconstrição cerebral reversível (RCVS). Descrevemos o caso de uma mulher de 35 anos com história de enxaqueca recorrente e sinais neurológicos focais, incluindo hemiataxia direita. Ela tinha sido diagnosticada com a doença COVID-19 duas semanas antes. A neuroimagem revelou estenose multifocal das artérias da circulação posterior e dissecção da artéria cerebelosa superior direita. A doente melhorou significativamente durante a sua hospitalização e manteve apenas ligeira ataxia. A interação entre a doença COVID-19, RCVS, e a dissecção arterial requer mais investigação.

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Introduction

Reversible cerebral vasoconstriction syndrome (RCVS) occurs in the setting of multifocal reversible vasospasm of the intracerebral arteries and manifests with recurrent thunderclap headache, focal neurological signs, seizures, and both ischemic and hemorrhagic strokes. It is commonly triggered by vasoconstrictive drugs or the post-partum state. Vascular abnormalities, such as cervical artery dissection, have been found in 7% to 12% of RCVS patients. To our knowledge, this is the first case reported of RCVS associated with dissection of the superior cerebellar artery (SCA) in a patient who recently recovered from COVID-19.

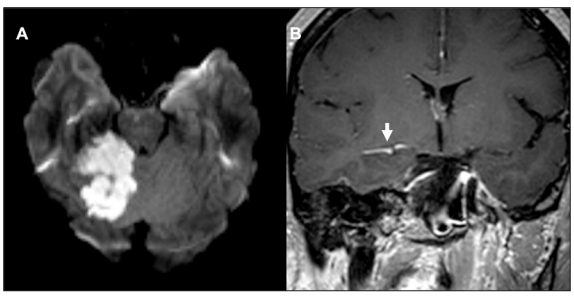

Figure 1. A: Axial diffusion-weighted magnetic resonance imaging (DWI) demonstrating right cerebellar hemisphere restricted diffusion consistent with acute cerebellar ischemia. B: Coronal oblique T1 weighted magnetic resonance vessel wall imaging (VWI) after gadolinium administration showed a long SCA hyperintense filling compatible with arterial dissection and intramural thrombus (white arrow).

Case Report

A 35-year-old woman presented to our emergency department with two episodes of bilateral occipital thunderclap headache separated by a three-hour interval, associated with nausea and vomiting, photophobia, and phonophobia. The pain intensity was rated with a 10 out of 10 intensity and did not improve after taking ibuprofen. She did not tolerate orthostatic position, straining, and lifting heavy objects. The patient denied fever, visual disturbances, confusion, trauma, and seizures.

Her past medical conditions included episodic migraine with typical aura, obesity, and lower limb varicose veins. She had had a positive nasopharyngeal swab for SARS-CoV-2 two weeks earlier, but no symptoms were present during the examination. She has been an active smoker since the age of 17 (18 pack-years) but denies alcohol or illicit drug use. Her current medications included sumatriptan *pro re nata* (PRN; 'as needed') and a progestin contraceptive implant, although she had not taken sumatriptan for this headache episode. Family history was irrelevant and vital signs were within normal parameters. Neurological examination revealed lethargy, gaze-evoked nystagmus, horizontal hypermetric saccades, left facial hypoesthesia and mild facial palsy, dysarthria, and right-sided limb ataxia. There was no hemiparesis, and visual fields were normal to confrontation. The patient had normal strength and reflexes of the upper and lower extremities bilaterally. There was no neck stiffness or other signs of meningeal irritation. The patient scored 6 points on the National Institutes of Health Stroke Scale.[1]

Initial laboratory studies including blood count, C-reactive protein, cardiac markers, and coagulation studies were unremarkable. Brain computed tomography (CT) was normal at admission but repeat brain CT 8 hours later showed decreased attenuation of the right cerebellar hemisphere which was in accordance with the occlusion of the proximal segment of the right SCA found on subsequent CT angiography. Transcranial Doppler (TCD) ultrasound showed increased velocity at the M1 segment of the left middle cerebral artery, A1 portion of the anterior cerebral artery, basilar artery, and both posterior cerebral arteries. Magnetic resonance (MR) angiography disclosed stenosis of the prepontine segment and fusiform dilatation of the ambient and quadrigeminal segments of the right SCA, with an eccentric T1 weighted hyperintensity suggestive of arterial dissection with intramural thrombus **(Fig. 1B)**. Luminal irregularities in the V4 segment of both vertebral arteries, basilar artery trunk, anterior inferior cerebellar arteries, left SCA, posterior cerebral arteries, A2 segment of both anterior cerebral arteries, and M2 branches of middle cerebral arteries were also found **(Fig. 2)**. Lumbar puncture, further bloodwork, including autoimmun-

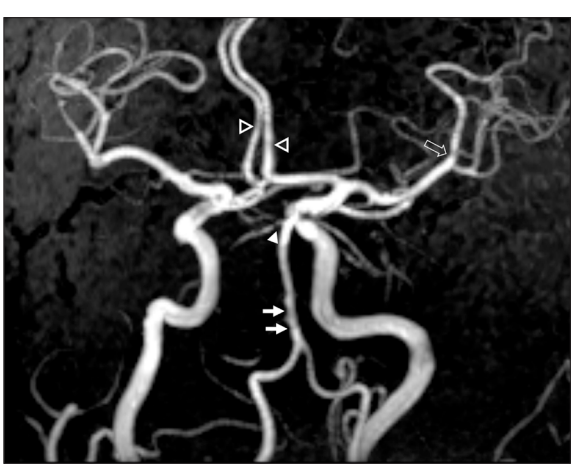

Figure 2. Maximum intensity projection (MIP) of magnetic resonance (MR) angiography showing stenosis of the prepontine segment of the right superior cerebellar artery (white arrowhead) followed by fusiform dilatation of the arterial lumen. Note the discrete multifocal luminal irregularities affecting the basilar artery (white arrows), the M1 segment of the left middle cerebral artery (empty arrow), and the A2 portion of both anterior cerebral arteries (empty arrowheads).

ity serum markers, and transthoracic echocardiogram, were unremarkable.

The diagnosis of RCVS was established (RCVS2 score of 7), and the patient was placed on nimodipine 30 mg three times daily orally, which was titrated to half of the dosage on day 7 of treatment given the improvements in cerebral blood flow velocities after repeating TCD. The patient received antiplatelet therapy during the hospital stay and was discharged to a rehabilitation facility with only mild right-sided ataxia and dysarthria.

Discussion

This case report describes a rare site for arterial dissection in a patient with the classic presentation of RCVS that had been previously diagnosed with SARS-CoV-2 infection. Thunderclap headache in RCVS mimic those of an aneurysmal subarachnoid hemorrhage. They are triggered by exertion or the Valsalva maneuver and recur in approximately 90% of the patients.[1] However, these headaches are different from previous migraine episodes. The mechanisms behind RCVS are not fully understood, but sympathetic-mediated changes in the vascular tone of the distal brain vasculature are generally accepted. Pregnancy, use of vasoconstrictive drugs, invasive neurosurgical procedures, unruptured saccular aneurysms, and cerebral venous thrombosis are commonly associated triggers.[2] MR angiography is the first-line non-invasive investigation and TCD may help predict ischemic complications.[3] RCVS may be complicated by subarachnoid hemorrhage, stroke, and arterial dissection. While the presentation of our patient may be typical, the dissection of the SCA associated with RCSV is an unusual finding. A series of 20 cases of RCVS associated with cervical artery dissection found that recurrent thunderclap headache was the most common presentation. The demographics of these patients matched those of RCVS and cervical artery dissection alone, although there was an unusually high frequency of multiple vertebral artery dissection. The lack of autonomic control of the posterior circulation in outcoming rapid changes in the blood pressure compared to the anterior circulation may explain why these arteries were predominantly involved when the dissection was multifocal.[4] At 3 months, patients were discharged with a modified Rankin Scale (mRS) score of 0 and vasoconstriction had resolved in all patients. However, 9 out of 30 dissected arteries developed either residual stenosis or aneurysms.[5]

Superior cerebellar artery dissection is already a rare entity on its own with only a few cases previously described. Contrary to the SCA, the posterior inferior cerebellar artery (PICA) seems to be the most dissected cerebellar artery. Cerebellar artery dissection is more common on the proximal arterial segment and shows a young female predominance. Cerebellar ischemia is found in almost half of the patients and headache is the most common presenting symptom.[6] Whether RCVS or SCA dissection started first in this patient remains elusive. RCVS may raise the likelihood of arterial dissection by increasing intraluminal pressure after the stenosis, but the intima dissection can also release vasoactive substances capable of inducing vasospasm.[5] The low incidence of both SCA dissection and RCVS makes a causal relation more likely.

Vasoactive substance use is a known trigger factor for RCVS, but not for arterial dissection. However, inflammation is a known risk factor for both. Cases of common, internal carotid and vertebral arteries dissection in asymptomatic COVID-19 patients have been reported,[7-10] but no record of cerebellar arteries involvement is described to date. The viral infection is set to trigger an inflammatory response with consequent endothelial damage and intimal vessel disruption.[11] These patients exhibited elevated leukocyte counts and inflammatory markers, so the lack of these findings and the timing of our patients' spontaneous dissection raises questions on whether the SARS-CoV-2 infection may have played a part in her presentation. The role of COVID-19 disease on RCVS is also not straightforward. RCVS has been found in two patients with COVID-19 disease,[12,13] but this phenomenon was not attributed directly to the infection itself. SARS-CoV-2 enters cells via the ACE2-receptors and the downregulation of those proteins may lead to increased brain vessel hypertonia and promote cerebral vasospasm.[14]

Our study was limited due to the lack of follow-up imaging, but it was deemed unnecessary given the favorable clinical improvement. However, the interplay between SARS-CoV-2 infection, RCVS, and arterial dissection requires further investigation. ■

Data availability statement

The data that support the findings of this study are available from the corresponding author, TP, upon reasonable request.

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Contributorship Statement / Declaração de Contribuição

TP, RM, RC, BM, RSR: Design, writing, and final approval.
JR, RD, TB, PA, PC: Critical review with intellectual input and final approval.

Responsabilidades Éticas

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