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Case Report Open Access

Post-Partum Reversible Cerebral Vasoconstriction Syndrome Associated with Coronary Vasospasm

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Abstract

Objective: To report the first case of reversible cerebral vasoconstriction syndrome (RCVS) diagnosed when reversible cardiac wall motion akinesia was observed after the administration of atropine in a 28-year-old lady presenting 4 weeks post-partum with a 2-week history of thunderclap headache.

Case presentation: We present a case of a 28-year-old lady, who presented to the American University of Beirut Medical Center, with a history of migraine with visual auras and peri-partum pituitary hyperplasia, presenting 4 weeks postpartum with a 2-week history of thunderclap headache.

Results: RCVS comprises a group of diseases characterized by reversible focal segmental narrowing of cerebral vessels, usually accompanied by thunderclap headache and sometimes focal neurological deficits.

Conclusion: Several case reports and case series have described the association of this disorder with vascular manifestations outside the cerebral vasculature, including dissection of both external and internal branches of the carotid arteries, unruptured saccular berry aneurysms and fibromuscular dysplasia of the extracranial internal carotid artery (ICA). In this short report we describe a patient with reversible cerebral vasoconstriction associated with reversible cardiac wall abnormalities.

Keywords: Stroke; Vasospasm; Intracranial; Seizures; Coronary vasospasm; Post-partum period

Abbreviations: DSA: Digital Subtraction Angiography; CAT: Computed Axial Tomography; MRI: Magnetic Resonance Imaging; MRA: Magnetic Resonance Angiography; ADC: Apparent Diffusion Coefficient; WMA: Wall Motion Abnormality; FLAIR: Fluid Attenuated Inversion Recovery; TEE: Trans-Esophageal Echocardiogram

Introduction

Reversible cerebral vasoconstriction syndrome (RCVS) comprises a group of diseases characterized by reversible focal segmental narrowing of cerebral vessels, usually accompanied by a thunderclap headache and occasional focal neurological deficits [1]. Several case reports and case series have described the association of this disorder with vascular manifestations outside the cerebral vasculature including dissection of both external and internal branches of the carotid arteries, unruptured saccular berry aneurysms, and fibromuscular dysplasia of the extracranial internal carotid artery (ICA) [2-4]. A case series published in 2014 described reversible cardiac Wall motion abnormalities (WMAs) and hypokinesis in 3 women with RCVS, 2 of whom were post-partum [3].

Case Presentation

We present a case of a 28-year-old lady, who presented to the American University of Beirut Medical Center, with a history of migraine with visual auras and peri-partum pituitary hyperplasia, presenting 4 weeks postpartum with a 2-week history of thunderclap headache. The patient's headache started acutely in maximal intensity in the occipital region and radiated all over the left side of her head along with blurry vision. She also experienced 4 generalized tonic-clonic seizures that occurred on the day of onset of her headache. Her neurological exam revealed bilateral peripheral visual field loss, which was an old finding that she had secondary to her known history of pituitary hyperplasia but was non-focal otherwise. Computerized Axial Tomography (CAT) of the brain without contrast done in the ER showed a cortical-subcortical hypodensity involving the right occipital and right temporal lobes consistent with an infarct (Figure 1a). Faint white matter hyper densities were observed in this area consistent with hemorrhagic transformation. The patient was admitted and underwent a magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) of her brain which revealed a high fluid attenuated inversion recovery (FLAIR) signal involving mainly the cortex of the medial aspect of the right occipital lobe (Figure 1b), with some small areas of restriction on apparent diffusion coefficient (ADC), consistent with a subacute infarct. The intracranial and extracranial vessels were found to be normal on MRA (Figure 1c) and on digital subtraction angiography (DSA) (Figure 1d). Cerebro-spinal fluid (CSF) studies, an extensive hypercoagulable panel including factor V Leiden mutations and methylenetetrahydrofolate reductase gene mutations, electrocardiogram, and troponin levels were all within normal limits. A tentative diagnosis of reversible cerebral vasoconstriction was made, and she was treated with nimodipine for vasospasm, short course of high dose corticosteroids for her headache as well as valproic acid for seizures. During her hospital stay the patient was scheduled for a trans-esophageal echocardiogram (TEE) as part of the workup of her stroke and considering her previous history of pericarditis at age of 12. The TEE procedure was complicated by severe bradycardia, chest pain, and reversible anterior wall akinesia lasting for 10 minutes which occurred after administration of 1 mg atropine IV. Therefore, we present a case of Reversible cerebral vasoconstriction syndrome in a 28-year-old postpartum lady associated with reversible coronary vasospasm.

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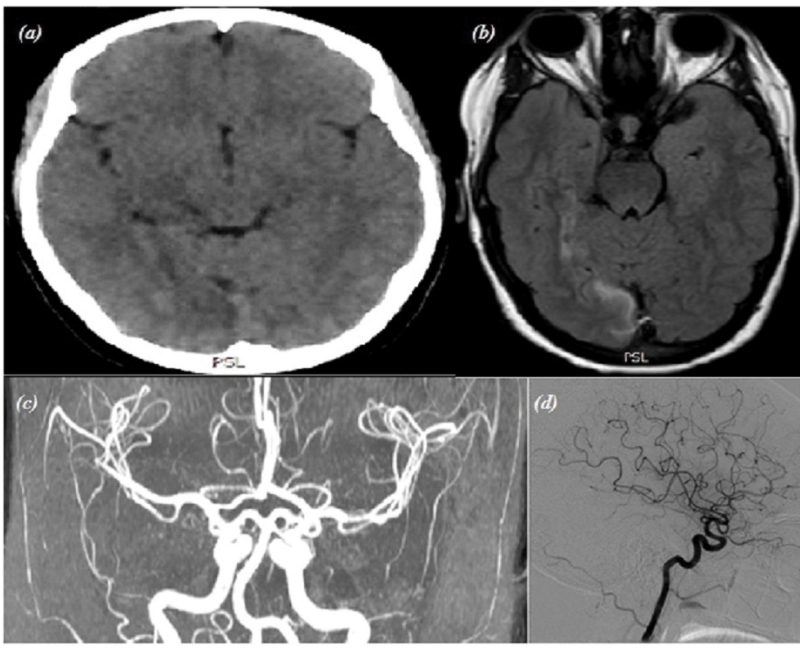

Figure 1: a) CT brain without contrast showing a cortical-subcortical hypodensity involving the right occipital and the right temporal lobes consistent with an ischemic infarct.

b) MRI FLAIR revealing a hyperintensity of the medial aspect of the right occipital lobe. Combined with the results of the ADC-MAP sequence (not shown), this suggested a sub-acute infarct.

c) MRA of the head showing normal intracranial and extra-cranial vasculature.

d) DSA cerebral vessels showing normal intracranial vessels.

Discussion

The leading hypothesis proposed by many to explain the association of RCVS with systemic vasoconstriction entails an alteration in vessel tone that leads to transient vasoconstriction [2]. Another hypothesis is that these patients have an abnormal vascular response to either intrinsic or extrinsic sympathomimetic agents [4]. Indeed, the post-partum state itself is considered a trigger for vasoconstriction, since it is postulated that the sudden decrease in the concentrations of estrogen and progesterone might trigger vasoconstriction [1,2]. In addition, several vasoactive drugs have been implicated as precipitants of this syndrome including selective serotonin re-uptake inhibitors (SSRIs), serotonin-norepinephrine reuptake inhibitors (SNRIs), cannabis, cocaine and triptans to name a few [1]. To our knowledge, atropine has not been reported as one of these precipitating drugs. However, due to its anti-muscarinic effects, it may be postulated that the resulting excess sympathomimetic activity triggered the coronary vasoconstriction in our patient.

Various diagnostic criteria have been suggested for RCVS, including the 2004 International Headache Society criteria and the criteria proposed by Calabrese et al. and Ducros et al. in 2007 [5,6]. Included in these criteria is the demonstration of reversible vasoconstriction by Magnetic Resonance Angiogram (MRA), Computed tomography angiography (CTA) (indirect) or conventional (direct) angiography by 12 weeks from symptom onset. The uniqueness of our case is attributable to the fact that the patient's presentation and evaluation were all suggestive of RCVS; however, multiple imaging studies (MRA, CTA and DSA) were normal. It was only after the reversible WMA was observed that this diagnosis was ascertained, unlike previously reported cases where cerebral vasoconstriction was observed before WMAs [3]. Normal angiography findings in RCVS were tackled in a 2007 prospective series by Ducros et al., who observed that the temporal pattern of occurrence of events, which progressed from hemorrhage in the first week to ischemia in the second week, suggests a centripetal progression of pathology within the cerebral arteries [6]. On clinical follow up 1 month later, the patient was completely symptom free.

Conclusion

Wall motion abnormalities have been previously observed in association with reversible cerebral vasoconstriction, but to our knowledge this is the first reported case of reversible wall motion akinesia following the administration of atropine. More studies on the pathophysiology of this rare disease and its association with extracranial vascular manifestations are needed, especially in the coronary vasculature which poses a serious concern in these patients.

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