

The Many Faces of Cerebrovascular Dysregulation: Reversible Cerebral Vasoconstriction Syndrome

Serebrovasküler Disregülasyonun Birçok Yüzü: Reversibl Serebral Vazokonstriksiyon Sendromu

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Dear Editor,

A 69-year-old man with a history of benign prostatic hyperplasia (BPH) admitted with the complaints of thunderclap headache, slurred speech and left-sided weakness. He reported having difficulty urinating due to BPH recently, he had to perform Valsalva maneuver several times.

On admission, neurological examination revealed neck stiffness and central facial paralysis, hemiparesis and extensor plantar response on the left side. Diffusion-weighted brain magnetic resonance imaging (MRI) showed multiple punctate diffusion restrictions. Susceptibility-weighted MRI images showed subarachnoid hemorrhage in the right frontoparietal convexity. There was a hyperintense signal change in the right occipital lobe on fluid attenuated inversion recovery (FLAIR) MRI images suggesting posterior reversible encephalopathy syndrome (PRES). An emergent cerebral digital subtraction angiography (DSA) revealed multifocal segmental vasoconstriction ("string and beads") of the petrous and the cavernous segments of the right internal carotid artery (ICA) and spasm of the middle and the anterior cerebral arteries on both sides. 1 mg intra-arterial nimodipine was selectively infused to each ICA. A control cerebral DSA obtained ten minutes after the nimodipine infusion showed marked improvement of vasoconstriction.

After the endovascular procedure, the patient was managed in neurointensive care unit to maintain optimal mean arterial pressure. He was given intravenous nimodipine (0.5 - 1 mg per hour) followed by oral nimodipine (60 mg every 4 hours). Brain perfusion MRI obtained

the day after the procedure did not show perfusion abnormality. A follow-up DSA obtained four days later showed normal luminal caliber of the intracranial vessels and no evidence of aneurysm or vascular malformation. The patient's clinical condition improved, and he was discharged after 14 days of hospitalization.

Reversible cerebral vasoconstriction syndrome (RCVS) is characterized by thunderclap headache and reversible multifocal constriction of cerebral arteries.¹ The most common precipitating factors are pregnancy, puerperium and vasoactive or illicit drug use; physical activities such as coughing, sexual intercourse and Valsalva maneuver have also been implicated.¹ We suggest that Valsalva maneuvers our patient performed several times to urinate might have caused RCVS.

The outcome of RCVS is usually favorable; however, complications such as ischemic stroke, convexity subarachnoid hemorrhage, PRES and intracerebral hemorrhage may occur.^{1,2} Differential diagnoses include aneurysmal subarachnoid hemorrhage, pituitary apoplexy, atherosclerotic and non-atherosclerotic intracranial vasculopathies and primary angiitis of the central nervous system.³ Although angiographic findings are highly sensitive, their specificity is low, and the diagnostic criteria of RCVS include the reversibility of angiographic findings within 12 weeks.⁴ Therefore, by definition, a definite diagnosis of RCVS can only be made retrospectively. To facilitate early diagnosis, intra-arterial nimodipine application has been proposed as a diagnostic tool.⁵ Linn et al.⁵ and Strunk et al.⁶ reported that vasoconstriction in all patients with RCVS completely resolved after intra-arterial nimodipine infusion. Nimodipine is also used for therapeutic purposes in RCVS (1).

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

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