A Storm in the Brain: Reversible Cerebral Vasoconstriction Syndrome Presenting as Intracerebral Hemorrhage - A Case Report

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Introduction

Reversible cerebrovascular syndrome (RCVS) is a rare and often underrecognized cerebrovascular condition characterized by sudden and severe headaches that occur over days to weeks due to underlying widespread reversible cerebral arterial vasoconstriction. Diagnosis of RCVS is supported by cerebral angiography, which reveals the classic "sausage on a string" appearance of constricted cerebral arteries, although initial neuroimaging might not show abnormalities. We present a case of a 52 year old female who presented to the hospital with new onset seizures and severe headaches and was found to have ICH. A diagnostic angiogram revealed a beading appearance of the anterior communicating artery (ACA) and middle communicating artery (MCA).

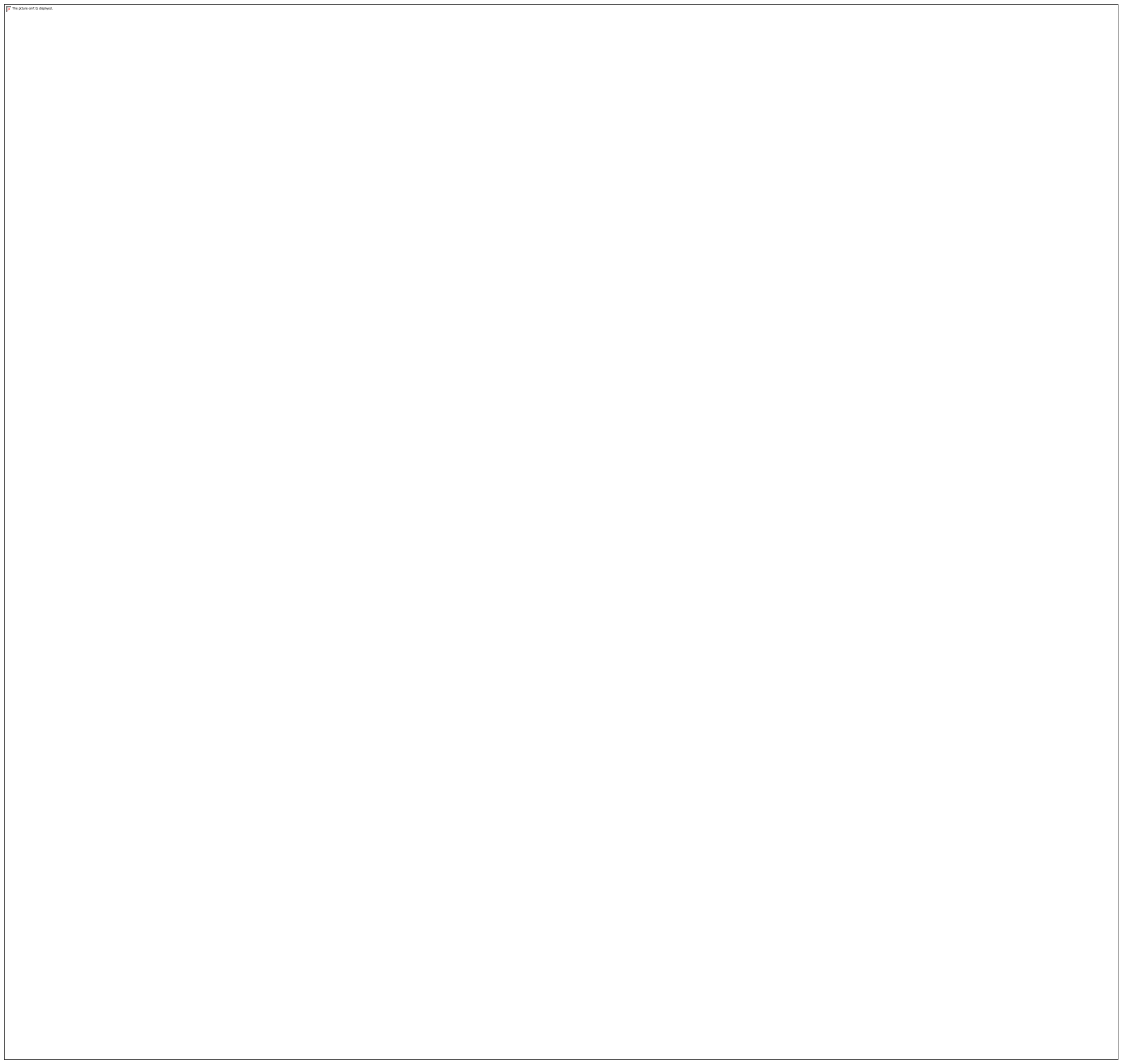
Case Presentation

Our patient is a 52-year-old female with a medical history of Deep Vein Thrombosis on Eliquis, Hashimoto's thyroiditis, peripheral neuropathy, cervical spine fusion surgery, migraines, and COPD initially presented with severe headaches, new onset seizures, and left-sided weakness. CT head showed acute right parietal intraparenchymal hemorrhage measuring 3.8 x 3.1 x 3.9 with surrounding vasogenic edema, with mass effect causing 4 mm leftward shift. Kcentra was given for Eliquis reversal. MRI of the brain showed stable right frontal intraparenchymal hemorrhage with surrounding vasogenic edema and 0.4 cm midline shift, and bilateral parieto-occipital white matter hypoattenuation concerning for Posterior reversible encephalopathy syndrome (PRES). Diagnostic cerebral angiogram revealed a beading

appearance of ACA and MCA, concerning vasculopathy. The immunological panel was notable for elevated IgM at 252, and the Vasculitis panel was negative for Anti SSA, SSB, C-ANCA, MPO, P ANCA, and atypical p-ANCA. Lupus PT/INR and PTT were elevated, consistent with lupus anticoagulants, though Cardiolipin, C3-C4, and B2 glycoprotein were negative. Lumbar puncture was not performed as MRI revealed edema and midline shift. Due to the high suspicion of RCVS, the patient was initiated on Nimodipine 60mg every 6 hr/day for 1 week, 60mg every 8 hrs/day for the next week, 60mg twice daily for the following week, and 60mg once a day for the last week, with improvement in her symptoms. The patient's home medication, Effexor, was reduced by 50%, given its vasoconstriction properties, and weaned off by the second week. The patient was scheduled for follow-up with Neurosurgery for a repeat angiogram in 2-3 months and MR vessel imaging.

Discussion and Conclusion

RCSV is a rare cerebrovascular condition that can mimic other vasculopathies with similar presentations, such as PRES or primary angiitis of the central nervous system. The exact pathophysiology of PRES is still not fully understood. Still, it is believed to involve dysregulation of cerebral autoregulation, referring to the brain's ability to maintain a consistent blood flow despite fluctuations in blood pressure by adjusting the constriction or dilation of cerebral blood vessels. Diagnosis is made mainly by excluding vasculopathy and by MR vessel imaging and cerebral angiogram with intra-arterial calcium channel blockers. This case indicates it is crucial to include RCVS in the differential diagnosis of sudden-onset headaches and seizures, as treatment is moderately benign. Therefore, early diagnosis is crucial to prevent complications of this reversible neurological disease.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