

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

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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 y/o female who presented to the hospital with new onset seizures and severe headaches and was found to have Intracerebral hemorrhage. 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 PMH of DVT on Eliquis, Hashimoto's thyroiditis, peripheral neuropathy, cervical spine fusion surgery, migraines, and COPD. She initially presented with severe headaches, new onset seizures, and left-sided weakness.

→ CT head 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. (**Image A**)

→ 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). (**Image B**)

Hospital Course

- 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-ANC.
- 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 Q6h for 1 week, 60mg Q8h the next week, 60mg BID for the following week, and 60mg QD 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.

Image A: MRI showing right frontal intraparenchymal hemorrhage with surrounding edema.

Image B: Diagnostic cerebral angiogram revealed a beading appearance of ACA and MCA

Image A

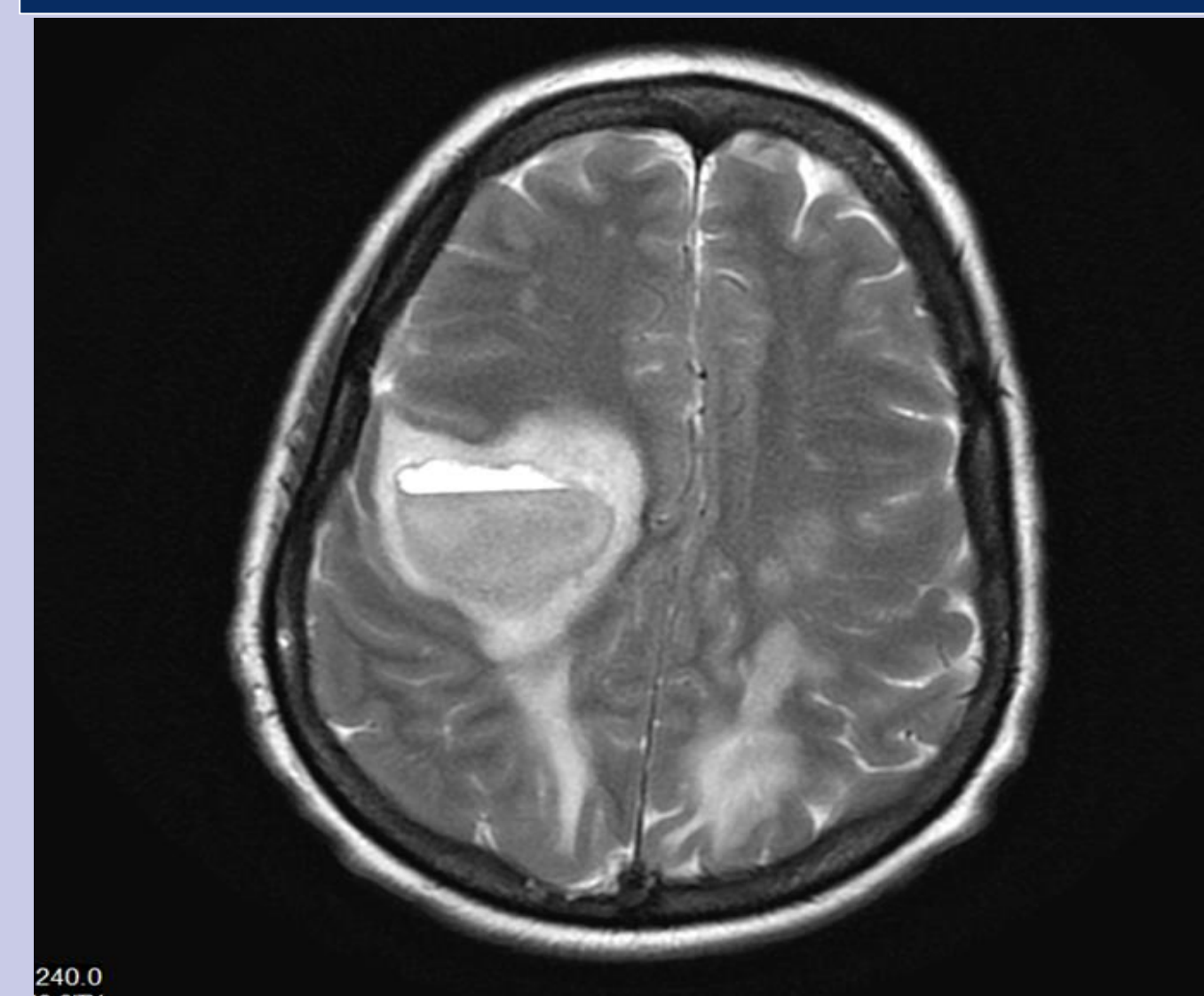
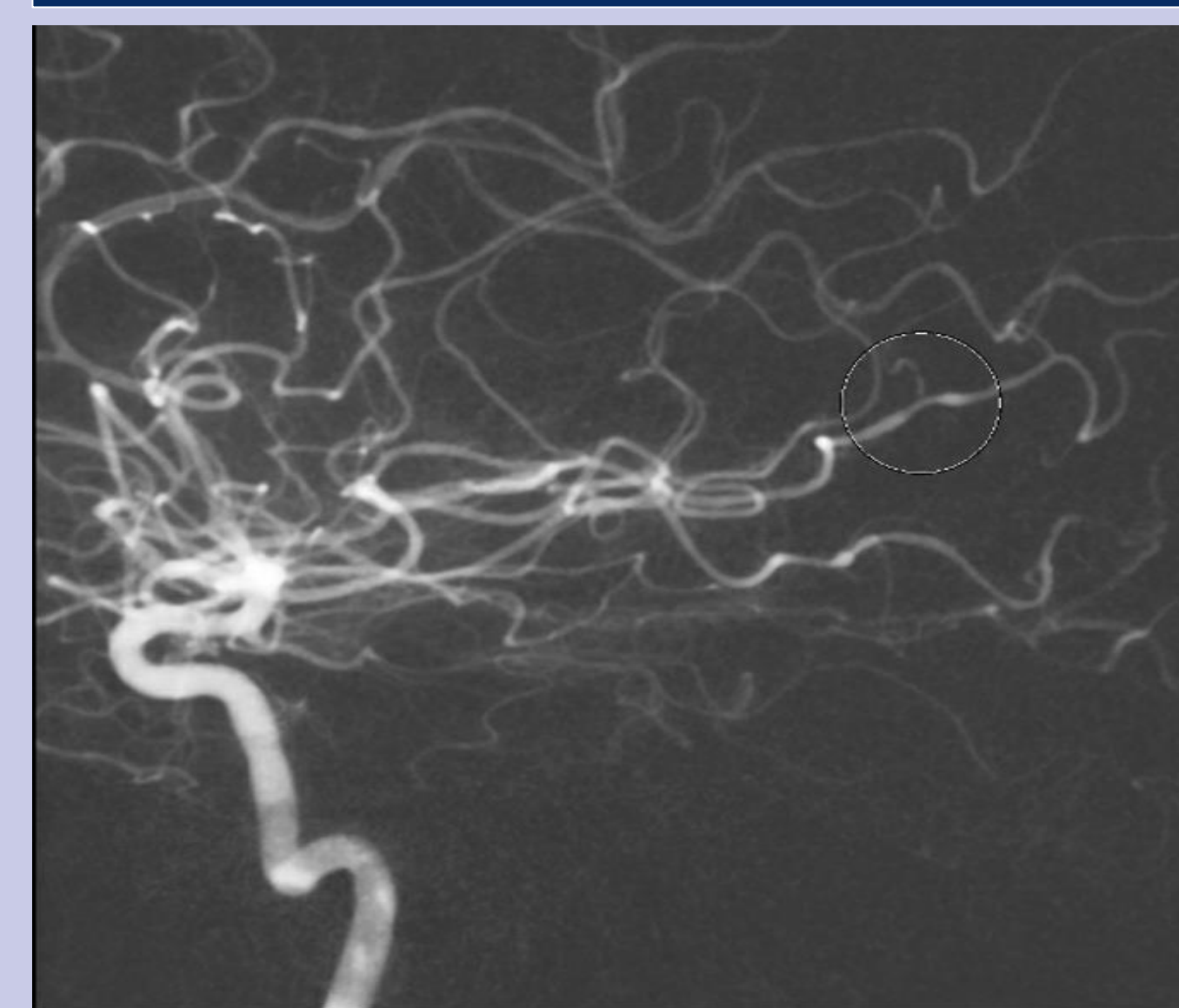


Image B



Discussion

- RCVS 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 RCVS 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 by clinical history, imaging, and by excluding vasculopathy. MR vessel imaging and cerebral angiogram with intra-arterial calcium-channel blockers can narrow down the differential.

Conclusion

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 essential to prevent complications of this reversible neurological disease.

References

