Primary CNS vasculitis with artery of Percheron and posterior circulation infarctions

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Case Study

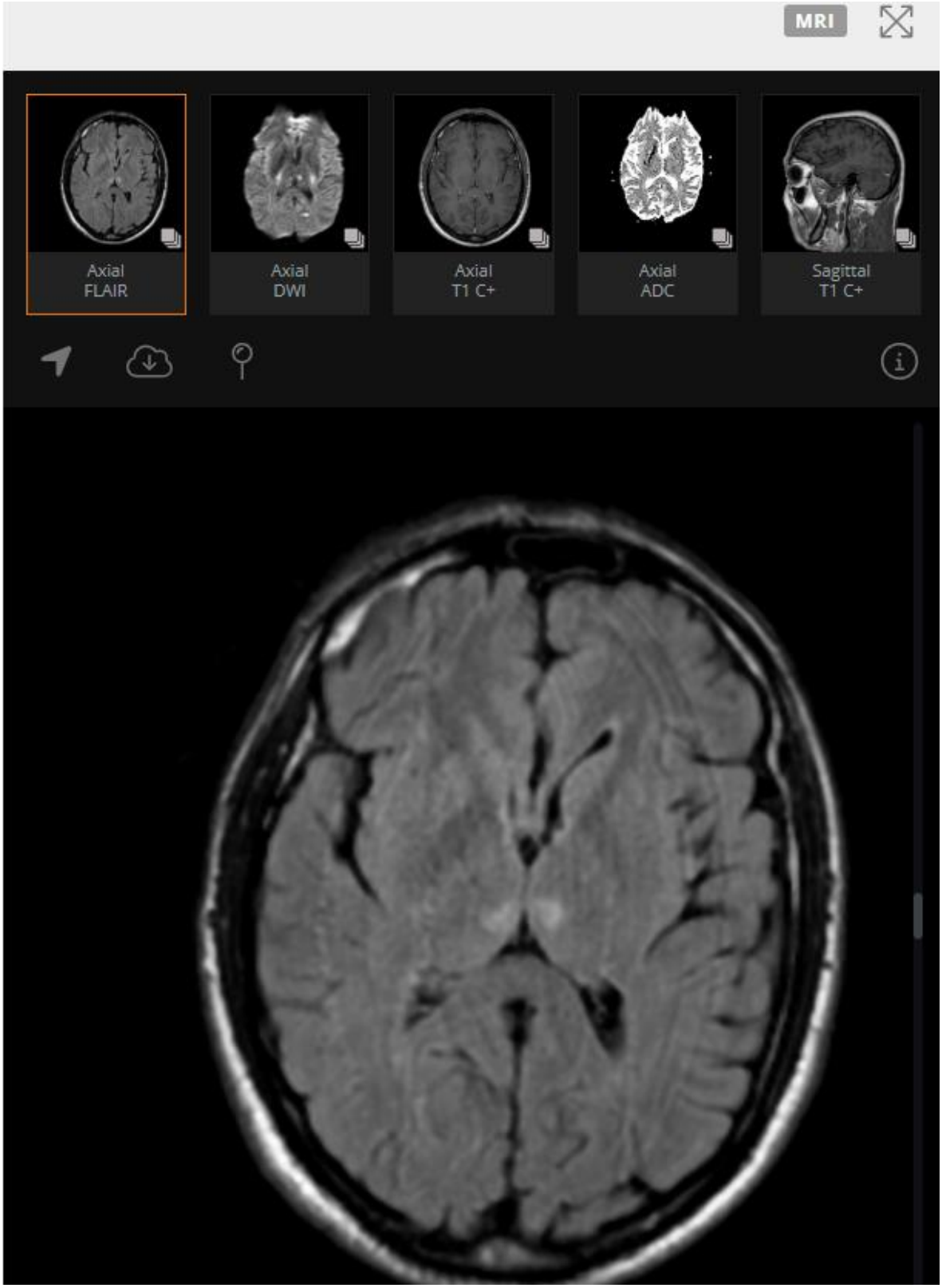
Presentation

The patient developed headache, dyscoordination, imbalance, memory problems, fever, and somnolence. Two days after manifestation, the patient was hospitalized with an altered mental state (GCS of 12). No previous history of atrial fibrillation, persistent headache, migraine, drug abuse, autoimmune, or systemic inflammatory disorders.

Patient Data

Age: 40 years

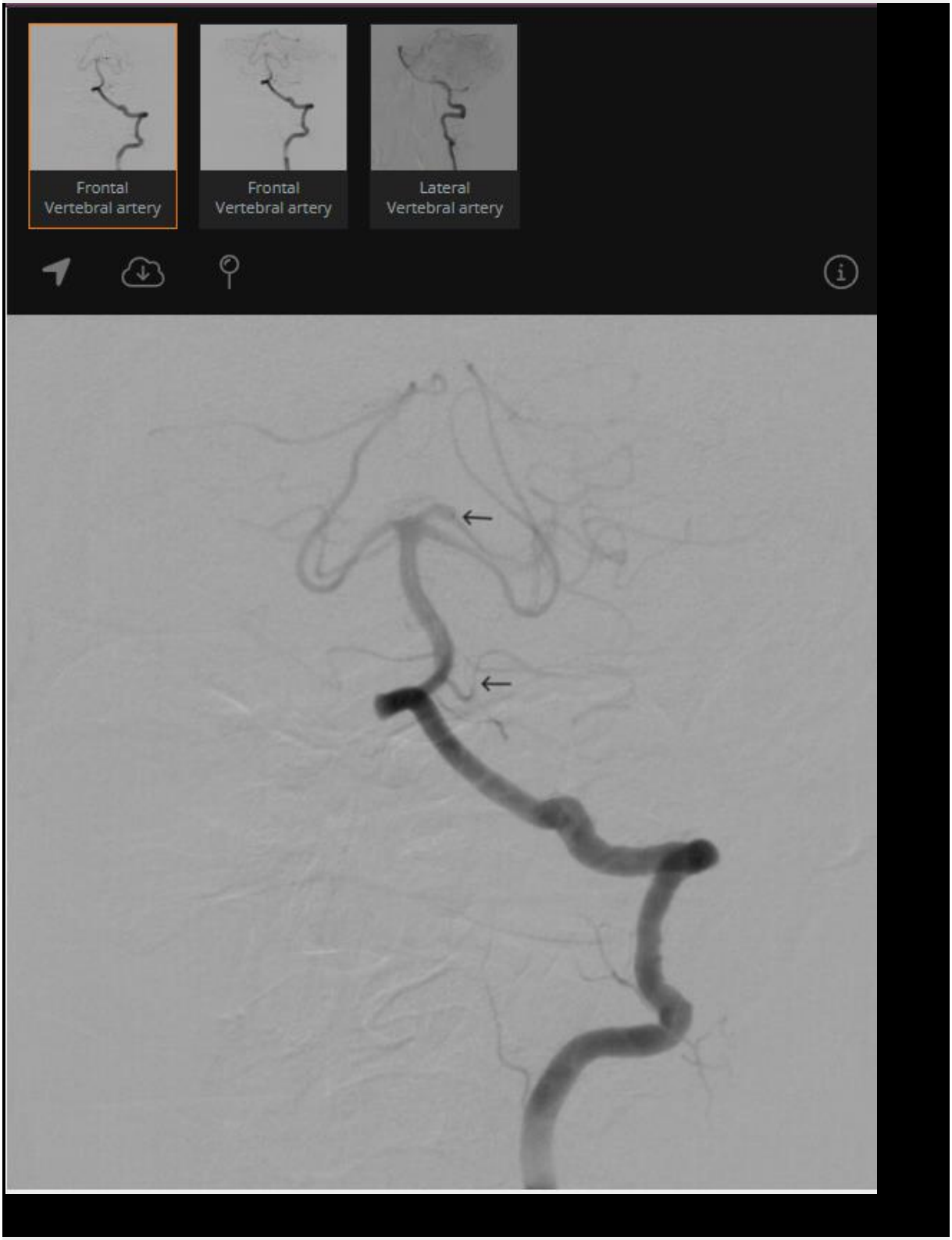
Gender: Male



Brain MRI demonstrates multiple T2 hyperintensities with diffusion restriction in the territories of the left superior cerebellar artery (SCA) and distal branches of both posterior inferior cerebellar artery (PICA) and left posterior cerebral artery (PCA). Bilateral paramedian thalamic and rostral midbrain T2 hyperintense lesions with diffusion restriction correspond to an occlusion of the artery of Percheron. No mass effect or contrast enhancement is observed in this lesions. There is multifocal leptomeningeal involvement. These changes are consistent with acute multifocal infarcts in the left occipital lobe, cerebellum, midbrain, and thalami.

Cerebral angiography

dsa

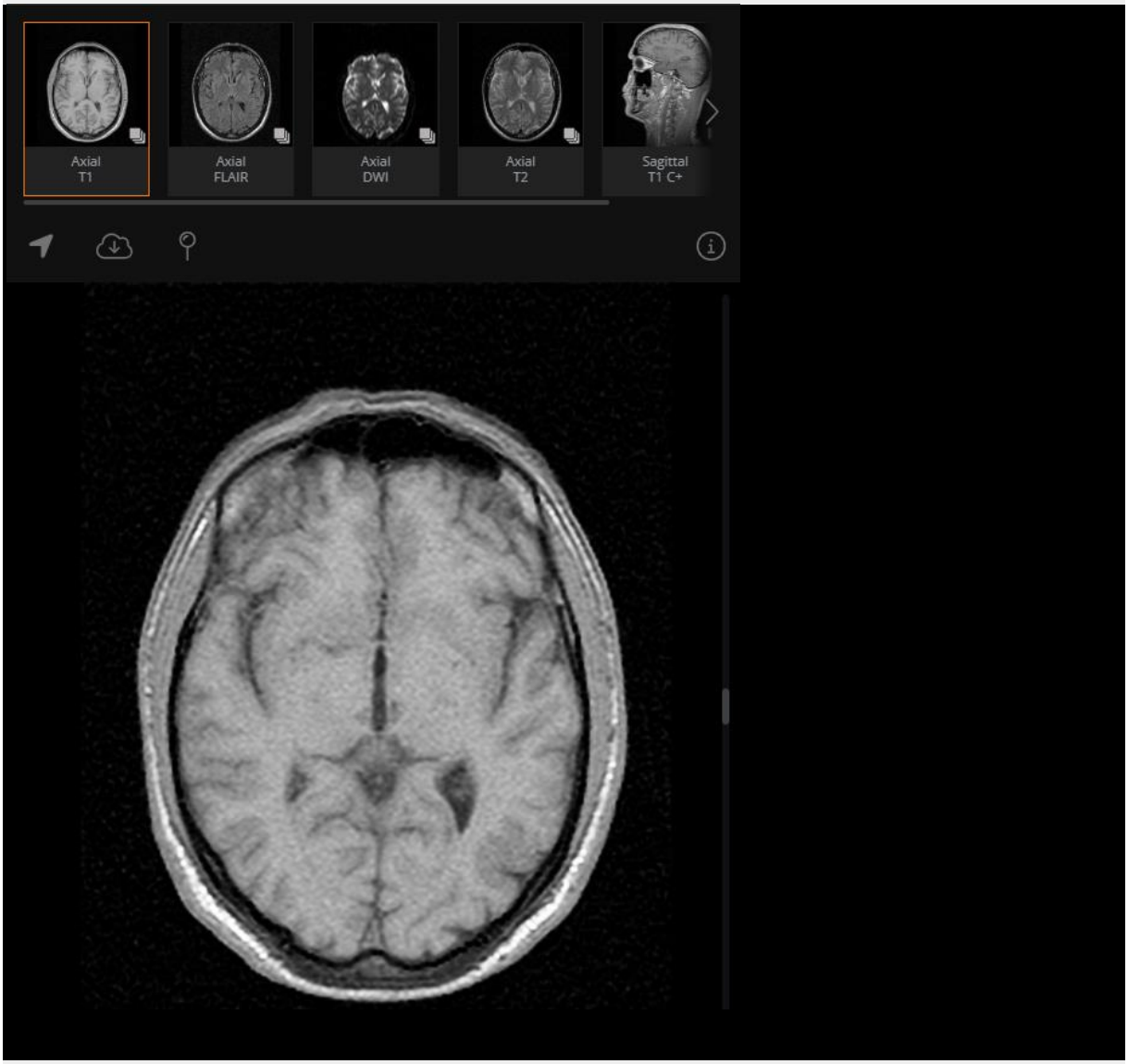


Cerebral angiography shows multifocal medium-small caliber vessel stenotic changes in the posterior circulation. The absence of the right posterior inferior cerebellar artery (PICA) is most likely due to

complete occlusion/high-grade stenosis at the outlet, along with decreased blood flow in the entire inferior part of the right cerebellar hemisphere. Remarkable long segment concentric stenosis of the left PICA, left superior cerebellar artery (SCA), and left posterior cerebral artery (PCA) is indicated with black arrows. Subtle blood flow in the stenotic artery of Percheron is visualized (red arrow). No significant vessel changes are observed in the anterior circulation. Both vertebral arteries and both internal carotid arteries are patent.

These findings, along with the brain MRI, represent multifocal ischemic strokes in the vertebrobasilar circulation due to long segment stenotic vessel changes, and the possibility of CNS vasculitis was considered.

6 months follow-up brain MRI



At the 6-month follow-up MRI, lacunes from previous strokes in the posterior brain circulation are observed. There is no evidence of new cerebral artery occlusions.

Case Discussion

CSF analysis revealed 10 WBC/μL with normal glucose and protein levels, no atypical cells or culture growth. No antibodies to TORCH infections or Borrelia were identified. CRP was elevated at 11.5 mg/dL, as well as ESR at 39 mm/hr, IgE at 1120.2 mU/L, and CD16+ NK cells in the blood, indicating inflammation. The rheumatologic workup was negative. Duplex ultrasound revealed bilateral resistive vertebral artery Doppler waveforms with no evident signs of atherosclerosis. Echocardiography was normal without valvular or septal pathology.

The combination of multifocal middle-small caliber vessel stenotic changes, inflammatory signs, and no specific autoimmune antibodies is suggestive of primary CNS vasculitis.[1]

The patient was initiated with a short-term course of corticosteroids, long-term therapy with immunosuppressive drug (chloroquine), pentoxifylline, and antiplatelet drugs. During the 1-year follow-up, he responded to the therapy with clinical improvement and a radiologically stable state.

The uniqueness of this clinical case is the coexistence of two clinically challenging disorders. The occlusion of the artery of Percheron, a rare anatomical variant of posterior cerebral perforator arteries [2], was caused by no less rare inflammatory neurological disease - primary CNS vasculitis.

References

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