Ischemic stroke with suspected intracranial vasculitis in antiphospholipid syndrome: a complex case of overlapping pathophysiology

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Background and purposes

Antiphospholipid syndrome (aPS) is an autoimmune prothrombotic disorder characterized by arterial, venous, and microvascular thromboses. While ischemic stroke is a common manifestation, the coexistence of intracranial large-vessel vasculitis is extremely rare in aPS and poses a diagnostic and therapeutic challenge. We present a complex case of ischemic stroke with radiological features suggestive of intracranial vasculitis in a patient with triple-positive aPS refractory to conventional anticoagulation.

Case report

A 50-year-old woman with a history of recurrent deep vein thromboses, pulmonary embolism, and prior stroke due to triple-positive aPS (treated with vitamin K antagonists) presented acutely with fluctuating right-sided paresthesia and mild hemiparesis. Her INR was 2.2 on admission. Urgent magnetic resonance imaging (MRI) revealed occlusion of the left intracranial internal carotid artery with **concentric wall enhancement** (*Figure 1 A,B*) suggestive of vasculitis and left hemisphere acute border-zone infarct. The patient rapidly developed a complete anterior circulation syndrome and underwent emergency endovascular therapy with intracranial carotid stenting. However, subacute **in-stent thrombosis** was observed on follow up imaging, and the device was removed. She was admitted to the ICU for further care and started on *intravenous methylprednisolone* and full-dose heparin. INR was adjusted to a higher therapeutic range (3.0–4.0). Repeat digital subtraction angiography (DSA) following further neurological deterioration showed **multifocal stenoses** (*Figure 1, C*) involving the bilateral anterior and middle cerebral arteries for which she was treated acutely with intra-arterial antiplatelet therapy. Due to disease progression despite optimal anticoagulation, the patient was started on *eculizumab* 900 mg weekly, targeting complement-mediated endothelial activation. This was followed by clinical stabilization and no further ischemic events during follow-up.

Neuroimaging

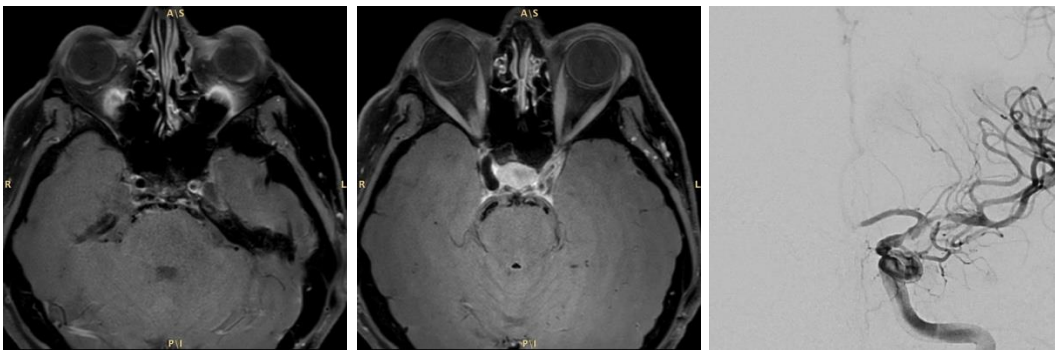

Figure 1. A, B) Brain MRI, T1 CE+, axial view. Note left intracranial internal carotid artery concentric vessel wall enhancement. **C)** Digital subtraction angiography showing multifocal (internal carotid, middle cerebral and anterior cerebral artery) stenosis.

Conclusions

This case highlights the importance of considering overlapping thrombotic and inflammatory mechanisms in aPS-related stroke, especially in the presence of radiological signs suggestive of vasculitis and poor response to standard anticoagulation. Complement inhibition has been used in catastrophic APS and APS-associated thrombotic microangiopathy, supported by case reports and preclinical data and is generally reserved for refractory or life-threatening cases due to limited clinical evidence. In selected cases, both immunosuppression and complement inhibition may be necessary to control both vascular inflammation and the prothrombotic state to prevent further ischemic events.