

Antiphospholipid Syndrome Presenting as Young Stroke with Diffuse ICA Narrowing in a 21-Year-Old Female: A Case Report

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Abstract:

Antiphospholipid Syndrome (APS) is an autoimmune prothrombotic disorder characterized by recurrent arterial or venous thromboses and/or pregnancy-related complications in the presence of antiphospholipid antibodies. We report the case of a 21-year-old female presenting with left hemiparesis and imaging findings suggestive of chronic infarct with diffuse right internal carotid artery (ICA) narrowing. Laboratory workup confirmed elevated antiphospholipid antibodies consistent with APS. This case highlights the importance of early recognition and evaluation of autoimmune prothrombotic states in young stroke patients.

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Introduction

Young stroke, defined as stroke occurring in individuals below 45 years of age, requires careful evaluation for uncommon etiologies such as autoimmune and thrombotic disorders. APS is a major yet under-recognized cause of arterial thrombosis in the young, with diagnosis relying on both clinical events and persistently elevated antiphospholipid antibodies, as defined by the revised Sydney criteria [1]. Stroke is the most frequent arterial manifestation of APS, accounting for nearly 20% of young stroke cases [2]. However, large-vessel involvement, particularly diffuse internal carotid artery (ICA) narrowing, is extremely rare, with only a few isolated reports in the literature [3,4]. This rarity makes the present case notable, as it emphasizes the importance of considering APS in the differential diagnosis when young patients present with atypical imaging findings suggestive of vasculopathy.

Case Presentation

A 21-year-old female, Miss Kaviya, presented with acute-onset left hemiparesis. At age 14, she was diagnosed with polyarthritis and treated with penicillin for several months. Two years later, she experienced right-sided weakness after a febrile illness with headache and hematemesis. The weakness improved over a week. On current admission, evaluation revealed elevated antiphospholipid antibodies. She was initiated on hydroxychloroquine, aspirin, and warfarin.

Neurological Examination

- Conscious, oriented

- Cranial nerves: Normal
- Tone and power: Left upper limb – reduced power with diffuse wasting
- Deep tendon reflexes: Right 2+, Left 3+
- Plantar response: Extensor on the left
- NIHSS score: 5/14

Investigations

MRI Brain: T2/FLAIR hyperintensity with gliotic changes in right parietal lobe; chronic infarct with ex vacuo dilatation of posterior horn of right lateral ventricle.

MRA: Thin flow in C2–C4 segment of right ICA; no flow in C5–C6 segment; MCA reformed via collaterals.

MRV: Hypoplastic left transverse sinus.

CT Angiogram: Diffuse luminal narrowing of entire right ICA without dissection or aneurysm.

Antiphospholipid Profile:

- Anti-β2 Glycoprotein I IgM: 32.66 (positive)
- Anti-β2 Glycoprotein I IgG: 2.05 (negative)
- Cardiolipin IgG: 1.90
- Cardiolipin IgM: 3.12
- Lupus Anticoagulant: Absent
- Protein C: 136%
- Protein S: 56%
- Factor V Leiden: Not detected
- Factor VIII: 110
- Homocysteine: 6.65 μmol/L

Diagnosis: Primary Antiphospholipid Antibody Syndrome with ischemic stroke due to diffuse right ICA involvement.

Treatment: Patient was initiated on dual antiplatelets, IV anticoagulation and statins and after optimization of INR discharged with single antiplatelet, warfarin, hydroxychloroquine and statins.

Review of Literature: APS is strongly associated with arterial events, particularly ischemic strokes in young individuals². Studies suggest that APS accounts for up to 20% of strokes in patients under 45 years [2]. While small- and medium-vessel thrombosis is well recognized, large-vessel involvement such as diffuse ICA narrowing is relatively rare but has been reported in isolated cases [3]. Cervera et al. reported stroke as the most frequent arterial manifestation of APS, with recurrence rates high in untreated patients [2]. Imaging in APS-related strokes may demonstrate both thrombotic occlusion and vasculopathy, mimicking atherosclerosis or dissection [4].

Hydroxychloroquine, beyond its immunomodulatory role, has been shown to exert antithrombotic and endothelial-protective effects in APS, and is increasingly recommended as adjunctive therapy with long-term anticoagulation [5]. Early recognition and prompt treatment are crucial, as the recurrence risk without therapy is substantial [6].

Discussion

This case highlights APS as a rare cause of large-vessel occlusion in young individuals. Diffuse ICA narrowing without dissection or atherosclerosis suggests autoimmune-mediated vasculopathy rather than conventional vascular pathology. High titers of anti- β_2 glycoprotein I IgM confirmed the diagnosis. Early anticoagulation remains the cornerstone of treatment to prevent recurrent ischemic events. Adjunctive therapies such as hydroxychloroquine may further improve vascular outcomes and are supported by recent meta-analyses [5].

This emphasizes the importance of suspecting APS in atypical stroke presentations in young patients.

Conclusion

APS should be considered in the differential diagnosis of young stroke patients, particularly when imaging reveals large-vessel narrowing without typical atherosclerotic features. Anticoagulation remains the mainstay of therapy, while early recognition is vital to prevent recurrent, disabling cerebrovascular events [1,2,6].

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