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Bilateral Internal Carotid Artery Occlusion Associated with the Antiphospholipid Antibody Syndrome

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Key Words
Carotid artery occlusion · Antiphospholipid antibody syndrome · Ischemic stroke · Cerebral arteriopathy

Abstract
A 39-year-old woman presented with a right-hemispheric stroke 1 year after she had suffered a left-hemispheric stroke. Her diagnostic workup was notable for bilateral occlusions of the internal carotid arteries at their origins and a positive lupus anticoagulant antibody test. There was no evidence of carotid dissection or another identifiable cause for her carotid occlusions. These findings suggest that the antiphospholipid antibody syndrome may be implicated in the pathological changes that resulted in occlusions of the extracranial internal carotid arteries. Young stroke patients who present with unexplained internal carotid artery occlusions may benefit from testing for the presence of antiphospholipid antibodies.

Introduction

Antiphospholipid antibodies are circulating immunoglobulins that are associated with both venous and arterial thrombotic events. The antiphospholipid antibody syndrome is defined by 2 major criteria: the occurrence of at least 1 clinical thrombotic event or pregnancy morbidity (either 1 otherwise unexplained fetal death after 10 weeks' gestation, 1 premature birth before 34 weeks' gestation because of preeclampsia, eclampsia or placental insufficiency, or 3 unexplained pregnancy losses before 10 weeks' gestation), and the

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presence of antiphospholipid antibodies measured on 2 or more occasions at least 12 weeks apart [1]. Venous thrombosis is the most common systemic manifestation of the antiphospholipid antibody syndrome, but within the central nervous system, arterial thrombosis is more common than venous thrombosis. Arterial stroke is the presenting symptom in 13% of patients with the antiphospholipid antibody syndrome [2, 3].

Imaging studies of patients with the antiphospholipid antibody syndrome have emphasized the parenchymal changes noted on CT and MRI [4]. These studies have shown that subcortical infarcts and hyperintense white matter foci are common in patients presenting both with and without clinical strokes. Fewer studies have reported on the angiographic findings in stroke patients with the antiphospholipid antibody syndrome. These studies have shown predominantly intracranial stem or branch occlusions and irregularities of the cerebral vessel walls consistent with vasculopathy [4, 5]. These angiographic findings suggest a link between the antiphospholipid antibody syndrome and cerebral arteriopathies and raise the possibility that arteriopathies and a hypercoagulable state are both implicated in the increased risk of arterial stroke. Whereas most arteriopathies in stroke patients with the antiphospholipid antibody syndrome involve the intracranial circulation, occasionally the extracranial internal carotid artery is affected [6]. Little is known about this manifestation and about the natural progression of stenosis of the extracranial internal carotid artery in patients with the antiphospholipid antibody syndrome. We describe a young patient who experienced progressive steno-occlusive disease of her bilateral extracranial internal carotid arteries in the setting of antiphospholipid antibodies.

Case Report

A 39-year-old woman was transferred to our hospital after being found unconscious at home. She was confused, but was able to give a history of left-sided weakness with decreased use of her left hand and difficulty bearing weight on her left leg that had begun abruptly 3 days prior to presentation.

She had had one prior outside hospital admission 1 year earlier for a left middle cerebral artery stroke that presented with expressive aphasia and right arm and leg weakness. Limited MRI at that time revealed 2 areas of restricted diffusion in the left parietal lobe with increased FLAIR signal and associated cortical/subcortical enhancement. Axial T1-weighted images showed the absence of a flow void in the left internal carotid artery, while the right internal carotid artery appeared patent based on the presence of a normal flow void. Imaging showed no evidence of dissection. The patient underwent stereotactic biopsy of the left parietal lesion to rule out a neoplasm. The results showed nonspecific necrotic changes and no evidence of a neoplasm. She was discharged to inpatient rehabilitation, and over the subsequent months her speech and motor function completely recovered.

Three years prior to her current presentation, the patient had had a miscarriage at 12 weeks' gestation. Her history was also significant for intravenous methamphetamine use, ending 13 years prior to presentation, and an 18 pack-year smoking history. She had no history of head or neck trauma. She did not have hypertension, diabetes, or hyperlipidemia. She did not use birth control pills and did not have a history of deep venous thrombosis or pulmonary embolism. Her family history was remarkable for systemic lupus erythematosus in her father and a brother who was diagnosed with thrombotic thrombocytopenia purpura.

On examination, her blood pressure was 126/80 mm Hg, and her heart rate was 91 beats/min and regular. There was no cardiac murmur. She was alert and oriented to person, but not place, time, or situation. She was able to follow simple commands, but was slow to

respond. Her speech was fluent, with intact repetition and naming and without aphasia or dysarthria. Cranial nerve examination was significant for a left homonymous hemianopsia and left lower facial weakness. Her motor exam revealed left hemiparesis, left hyperreflexia, and a left Babinski sign. She had intact sensation of light touch bilaterally, but extinction on the left with double simultaneous stimuli. Movements were slow on the left side, but there was no ataxia. Gait was not assessed.

Abnormal coagulation studies included a prolonged activated partial thromboplastin time at 42.6 s (normal range 25.1–37.6 s) which did not correct with a 1:1 mix with normal plasma, a thromboplastin inhibition test that was strongly positive for lupus anticoagulant, and a dilute Russell's viper venom test that was prolonged at 64.3 s (normal 29–46 s) which corrected with the addition of phospholipids. Other coagulation studies, which included anticardiolipin and anti-β2 glycoprotein antibodies, were within normal limits. Erythrocyte sedimentation rate (55 mm/h) and C-reactive protein (5.7 mg/l) were moderately elevated, and the antinuclear antibody assay was elevated at 1:160. Anti-dsDNA was negative. Other studies were normal or negative, including complete blood count, platelet count, renal, liver, and thyroid functions, hemoglobin A1c, and LDL cholesterol. Electrocardiogram and transthoracic echocardiogram were normal.

A CT scan showed areas of evolving infarction in the right anterior and middle cerebral artery distributions. CT angiography showed bilateral internal carotid artery occlusions extending from the level of the bifurcation to the supraclinoid segments. MRI showed areas of acute infarction within the right middle cerebral artery-anterior cerebral artery watershed territory and chronic infarcts in the left middle cerebral artery-anterior cerebral artery watershed territory (fig. 1).

Conventional angiography confirmed bilateral internal carotid artery occlusions in the proximal cervical segments, with reconstitution of the carotid arteries at the supraclinoid segments via the ethmoidal branches of the distal internal maxillary arteries and the ophthalmic arteries (fig. 1). Posterior communicating arteries were present bilaterally. The posterior circulation was normal, with extensive collaterals to the anterior circulation via the posterior communicating arteries. No abnormalities concerning for vasculopathy or dissection were noted in the aortic arch or in the carotid, vertebral, subclavian, renal, or femoral arteries.

Discussion

Our patient met clinical and laboratory criteria for a diagnosis of the antiphospholipid antibody syndrome. She was young, had a characteristic history of prior spontaneous abortion at more than 10 weeks' gestation, and had a strongly positive thromboplastin inhibitor test consistent with lupus anticoagulant antibodies. Her presentation with strokes secondary to bilateral carotid artery occlusions suggests an association between the antiphospholipid antibody syndrome and a vasculopathy involving the extracranial segments of the internal carotid arteries.

The causal relationship between the antiphospholipid antibody syndrome and bilateral carotid occlusions is further supported by the absence of reasonable alternative causes of carotid occlusions in this young patient. Specifically, there was no history of head or neck trauma and no evidence of dissection or atherosclerosis on imaging. She had no evidence of a proximal embolic source. She was a smoker, but she had no history of hypertension, hyperlipidemia, or other vascular risk factors.

Her medical history indicates that vasculopathy of the internal carotid arteries in the setting of the antiphospholipid antibody syndrome can be rapidly progressive. She experienced a left middle cerebral artery stroke at the age of 38 years. MRI at that time was notable for an absence of a flow void in the left internal carotid artery, consistent with a left carotid occlusion, and the presence of a flow void in the right internal carotid artery, indicating patency of that vessel. Within 1 year, however, she presented with a right middle cerebral artery stroke, and workup revealed progression to bilateral carotid artery occlusions.

In summary, the findings suggest that the antiphospholipid antibody syndrome may be implicated in pathological changes that can result in occlusions of the proximal segments of the internal carotid arteries. Consequently, young stroke patients who present with unexplained internal carotid artery occlusions may benefit from testing for the presence of antiphospholipid antibodies.

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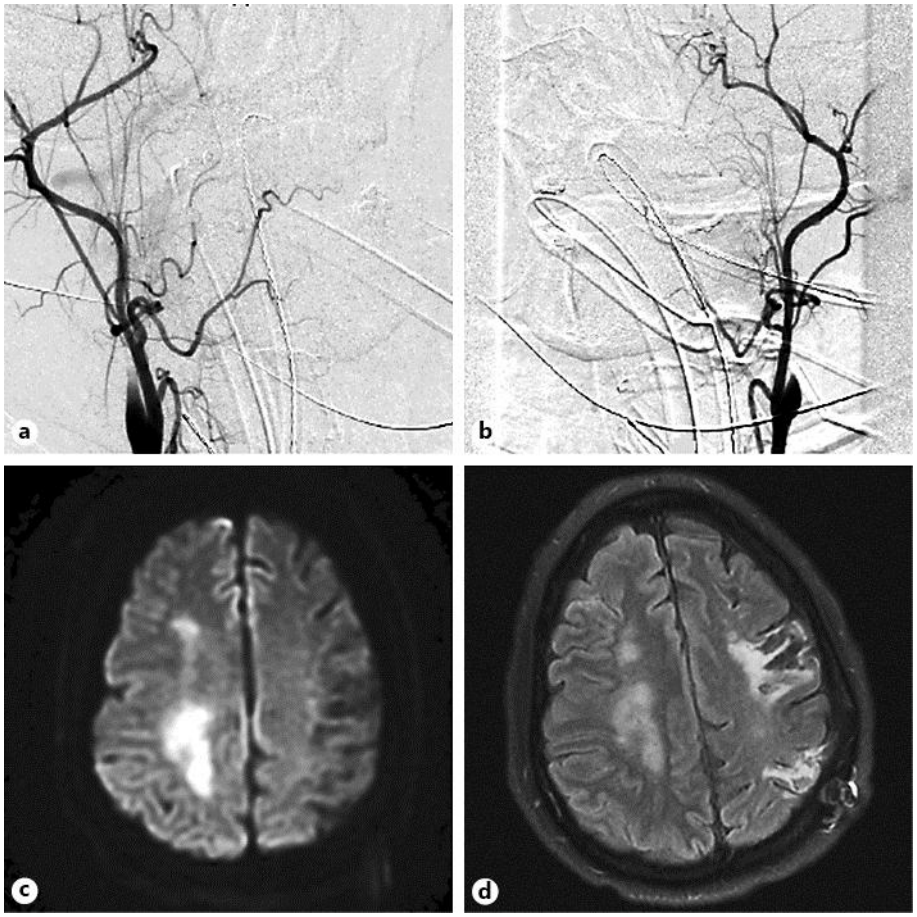

Fig. 1. Angiogram shows occlusions of the right (**a**) and left (**b**) internal carotid arteries in the proximal cervical segments. Infarction in the right middle cerebral artery-anterior cerebral artery watershed territory is seen on the DWI image (**c**). The T2 FLAIR image (**d**) shows the same area of infarction in the right middle cerebral artery-anterior cerebral artery watershed territory as well as regions of chronic infarction in the left middle cerebral artery territory.