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Case Reports and Series

Acute Limb-Threatening Ischemia Associated With Antiphospholipid Syndrome: A Report of Two Cases

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ABSTRACT

Acute limb ischemia results from sudden deterioration in the arterial supply to the limb, occasionally leading to limb loss or fatality. Antiphospholipid syndrome (APS) is known to induce acute limb ischemia among the various etiologies responsible for arterial obstruction. APS is a systemic autoimmune disorder characterized by a combination of arterial and/or venous thrombosis and limb loss. It is often accompanied by a mild-to-moderate thrombocytopenia and elevated titers of antiphospholipid antibodies, including the lupus anticoagulant and the anticardiolipin antibodies. In the present report, we present 2 cases of acute limb ischemia due to APS associated with systemic lupus erythematosus. Angiography revealed arterial obstruction distal to the popliteal artery in both patients, and each patient eventually underwent below-the-knee amputation. Surgeons treating acute limb ischemia should remember APS, although this disease might not be common in daily clinical practice.

Acute limb ischemia represents one of the toughest challenges that extremity surgeons encounter in their clinical practice. Its diagnosis and initial assessment are largely clinical, and a diagnostic error risks serious consequences to the patient, including amputation of the limb or even death. Acute limb ischemia is often the end-of-life condition that confronts a patient with multiple medical comorbidities. A careful clinical assessment is therefore very important for preservation of the limb and, in many cases, life salvage. Acute limb ischemia develops when an abrupt interruption of blood flow to an extremity occurs, usually because of either embolic or thrombotic vascular occlusion. Antiphospholipid syndrome (APS) is an autoimmune disease characterized by the clinical features of recurrent thrombosis in the venous and/or arterial circulation, often associated with loss of the fetus in a pregnant female (1). The Euro-Phospholipid project estimates that the manifestations of APS in the lower extremities include deep vein thrombosis in 38.9%, superficial thrombophlebitis in 11.7%, and arterial thrombosis in 4.3% (2). In the present report, we describe 2 cases of lower extremity arterial thrombosis associated with APS in patients with systemic lupus erythematosus (SLE).

Case Reports

Patient 1

A 48-year-old Japanese male diagnosed with SLE-associated APS had had his left leg scratched 2 months earlier, and he was referred to the Saga University Hospital for treatment of a refractory leg ulceration with cellulitis that had been present from February 2008 to August 2008. Three days before his referral, he had experienced sudden pain in his left limb and a high fever. He presented with widespread cutaneous necrosis (Fig. 1), and leg angiography was obtained. The angiogram revealed obstructions in both the anterior and the posterior tibial arteries of his left leg (Fig. 2). We identified the condition as acute limb ischemia with infection, with no indication for revascularization. Instead, we recommended major amputation for control of the pain and infection. After 14 days of medical stabilization and supportive therapy, the patient underwent surgery. We confirmed the presence of emboli in the anterior and posterior tibial arteries and venae comitantes during the operation (Fig. 3). Histopathologic examination disclosed organized thrombi, hypertrophy of the intima with crystal-form cholesterol, invasion of foam cells, and

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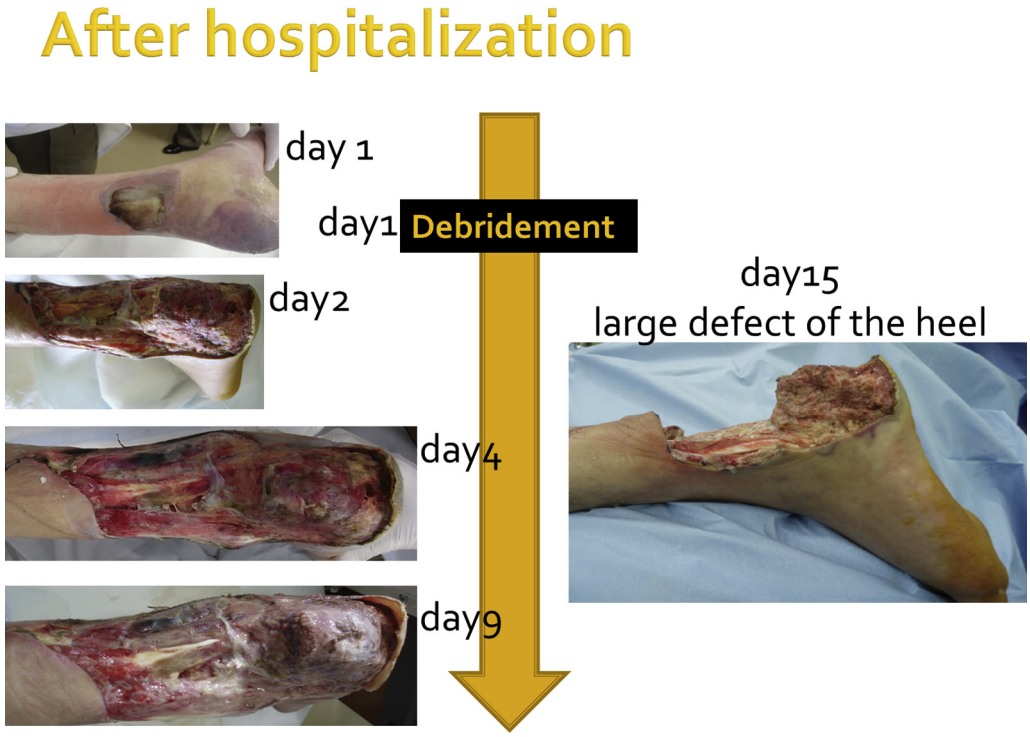

Fig. 1. View of patient 1 showing widespread cutaneous necrosis.

thromboarteritis (Fig. 4). After surgery, anticoagulant therapy was initiated with oral doses of warfarin 5 mg/day, aspirin 100 mg/day, and sarpogrelate 300 mg/day. Immunosuppressive therapy, under the guidance of the rheumatologist, was initiated with oral doses of prednisolone 35 mg/day, which was gradually decreased, and intermittent pulse intravenous cyclophosphamide therapy (750 mg/day). He survived the surgery and has been walking with appliances for >5 years since the amputation.

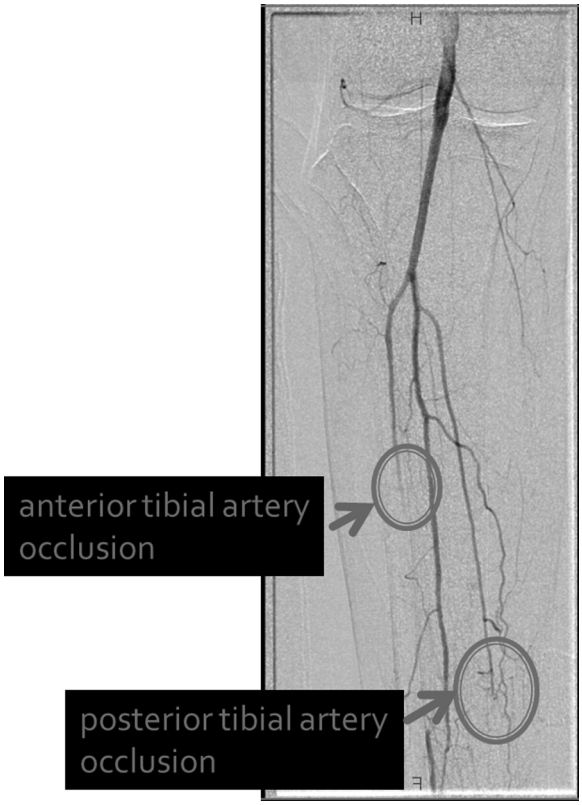

Fig. 2. Angiogram showing obstruction.

Case 2

A 65-year-old Japanese female presented with the sudden onset of gangrene of the right forefoot and heel in October 2012 (Fig. 5). She had 2 children and no history of a miscarriage. A contrast

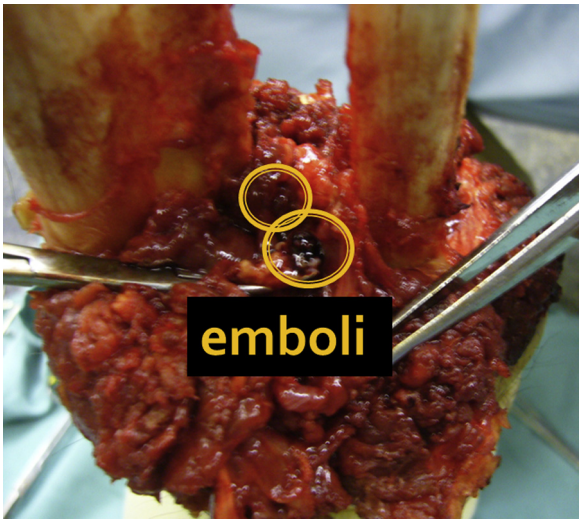

Fig. 3. The presence of emboli in the anterior and posterior tibial arteries and venae comitantes was confirmed during surgery.