Multiple Sterile Abscesses in Antiphospholipid Antibody Syndrome

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Antiphospholipid antibody syndrome (APS) is a systemic disorder characterized by elevated antiphospholipid antibodies and multiple-organ involvement. Reported cutaneous manifestations of APS include various skin lesions but not multiple sterile abscesses. We report a case of secondary APS with multiple sterile abscesses as a novel cutaneous manifestation. We believe these abscesses should be included among the cutaneous symptoms of APS.

Antiphospholipid antibody syndrome (APS) manifests as arterial or venous thrombosis, thrombocytopenia, and recurrent spontaneous abortion with laboratory abnormality of elevated antiphospholipid antibodies. Different types of skin lesion have been reported as cutaneous manifestations of APS.[1-8] In our case report, we describe a 21-year-old woman with multiple sterile abscesses on extremities in secondary APS with underlying systemic lupus erythematosus (SLE) and polymyositis. To our knowledge, these abscesses have not been reported in other patients.

Case Report

A 21-year-old woman had multiple painful swellings on the upper and lower extremities. Seventeen months before her visit, SLE and polymyositis were diagnosed based on general weakness; arthritis; myalgia; muscle weakness; and abnormal laboratory findings, including proteinuria, severe anemia (hemoglobin level, 5.3%), increased level of muscle enzymes (eg, creatinine phosphokinase, aldolase, lactate dehydrogenase), presence of antinuclear and anti–double-stranded DNA antibodies, and abnormal findings of muscle biopsy and electromyography.

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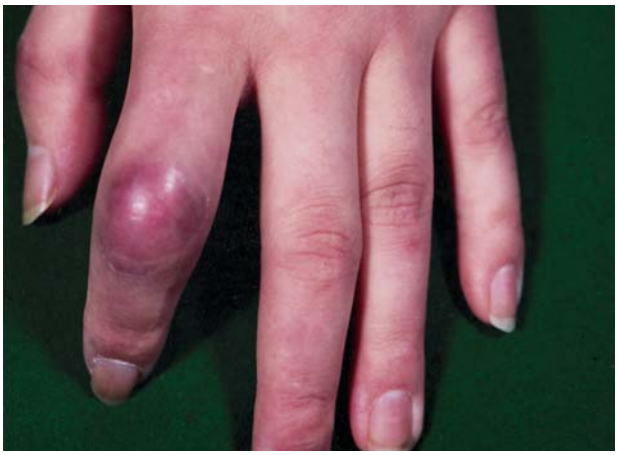

Figure 1. A purple abscesslike swelling on dorsum of the index finger.

Findings from a skin examination showed multiple erythematous, diffuse swellings on the upper extremity and both hands (Figure 1). Needle aspiration of the swellings showed that their content was a yellowish puslike clump. Repeated bacterial (aerobic and anaerobic) and fungal cultures and Gram staining with the abscess content did not show any evidence of microorganisms. There were diffuse, erythematous, mottled patches of livedo reticularis with acral cyanosis on both lower extremities (Figure 2). A biopsy specimen from the perilesional skin of the abscess showed endothelial proliferation, vascular thrombosis, and perivascular lymphocytic infiltration in the small vessels—compatible with the findings of lymphocytic vasculitis. Direct immunofluorescence showed C3 and fibrinogen along vascular walls in the upper dermis (Figure 3). A granular pattern of IgG also was evident in the dermoepidermal junction. IgG anticardiolipin antibody was positive (25.6 U/mL) in an enzyme-linked immunosorbent assay (normal, <15 U/mL).

Based on the diagnosis of secondary APS from underlying SLE and polymyositis, the patient was started on low-dose prednisolone orally. During the

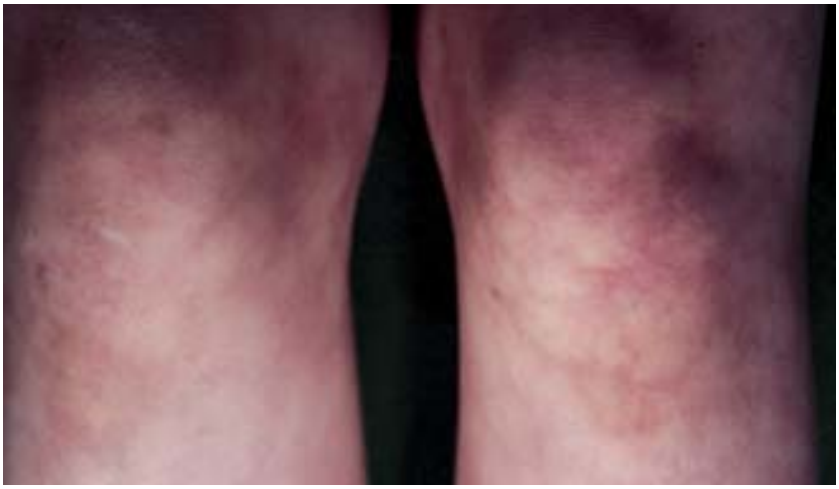

Figure 2. Diffuse, erythematous, mottled patches on lower extremities.

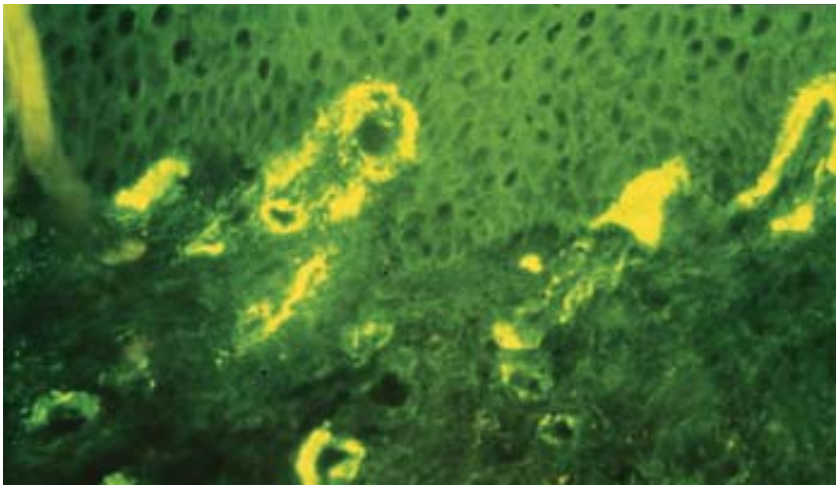

Figure 3. Deposition of fibrinogen around vascular walls in the upper dermis of perilesional skin (direct immunofluorescence, original magnification ×200).

5-day treatment, she had a sudden onset of neurologic symptoms—headache, nausea, vomiting, seizure, and homonymous hemianopsia. Computed tomography and magnetic resonance imaging of the brain showed multifocal ischemia and vascular abnormalities of the temporal and occipital lobes. Within 2 weeks of oral administration of prednisolone 60 mg per day and aspirin 300 mg per day, the abscesses resolved rapidly, and the neurologic symptoms went into complete remission.

Comment

Secondary APS is caused by many underlying diseases and is positive with circulating anticoagulant antibodies. The antibodies were first detected with SLE, but they have also been found in the presence of drugs, neoplasia, or infectious disease—or even without detectable underlying disease. In our case, SLE and polymyositis were found to be underlying causes of secondary APS.

Skin lesions are the first signs of APS in 41% of APS patients, and various cutaneous manifestations have been reported.[9] Therefore, dermatologists should be aware of the importance of cutaneous lesions in making an early diagnosis of APS. Livedo reticularis is the most important cutaneous finding of APS, probably because of the vascular occlusion.[10,11] Livedo reticularis was found in 23% to 48% of patients with SLE and was correlated to the increased level of antiphospholipid antibodies.[10,12] It also has been found in other autoimmune diseases, vasculitis, and hematologic conditions.[1] In particular, widespread livedo reticularis is an important cutaneous manifestation of Sneddon's syndrome, in which neurologic symptoms are associated with vascular abnormalities of the brain. Our patient manifested neurologic symptoms after cutaneous symptoms, which suggests widespread involvement of vasculopathy of the dermis and of the central nervous system. As with our case, patients with livedo reticularis and an increased level of IgG anticardiolipin antibodies had a higher incidence of cerebrovascular disease in SLE.[10] Cutaneous ulcers are another cutaneous finding in patients with APS, some of whom showed

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pyoderma gangrenosum–like lesions.[2] Cutaneous necroses,[3] including necrotizing purpura,[4] distal ischemia/gangrene,[2,5] thrombophlebitis,[2] hemorrhage,[2] and atrophie blanche (ie, porcelain-white scars),[5] also were reported in APS. In reviewing the literature, however, we found no reports of multiple sterile abscesses as a cutaneous manifestation of APS.

The mechanism for the production of sterile abscesses, as observed in our case, is unknown. We speculate that they are caused by severe vascular damage with serious circulatory disturbance—a pathogenesis similar to that of livedo reticularis but with higher degrees of severity.

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