Peripheral Gangrene in a Rare Scleroderma Overlap Syndrome

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A 48-year-old woman, a known case of systemic sclerosis and interstitial lung disease, presented with complaints of pain and blackish discoloration of the toes of both feet for 1 month (Figure 1). There was no history of fever, muscle pain, weakness, skin rash, joint pain or bleeding from any site. On examination, the patient had a pulse rate of 90/minute which was regular and the pulse volume was low in the left brachial and radial arteries. Dorsalis pedis pulse was not palpable on both sides. There was a difference of 30 mmHg between the right and the left arm in systolic blood pressures. Musculoskeletal examination showed auto-amputated fingers of both hands, telangectasia, calcinosis cutis and skin thickening (Figures 2 and 3). Dry gangrene was present in both feet (right > left). Investigations showed raised ESR and CRP and positive ANA and ant-centromere antibodies. Radiographs of both hands showedacro-osteolysis, erosions and joint space narrowing at proximal interphalangeal joints (Figure 4). Arterial Doppler of both lower limbs was reported as showing features of small vessel disease. CT aorta angiogram revealed focal stenosis of branches of the aorta which was suggestive of type V Takayasu arteritis (Figures 5, 6 and 7). Since our patient had progressive gangrene, an active vasculitic process was thought of. The patient was treated with pulse methylprednisolone followed by oralPrednisolone because of extensive gangrene in both feet. Since there was no improvement in the peripheral gangrene even after a month, she was given a cyclophosphamide pulse. However, the patient succumbed to a respiratory infection 3 weeks later.

Scleroderma is a debilitating autoimmune connective tissue disorder affecting the skin, lung, pulmonary artery, renal artery and the alimentary tract. Diffuse microangiopathy, inflammation due to autoimmunity, and visceral and vascular fibrosis are the cardinal pathophysiology features of scleroderma. Raynaud phenomenon is present in almost all patients with scleroderma. It can lead to digital pitting, ulceration and gangrene.¹

Overlap syndrome is a well-known entity among rheumatological diseases. Scleroderma overlap syndrome can present with other connective tissue disorders like polymyositis, systemic lupus erythematosus and rheumatoid arthritis etc.² Typical vasculitis with inflammatory infiltrates damaging blood vessels has rarely been reported in patients with Scleroderma.³

Takayasu arteritis is a chronic large-

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vessel arteritis that predominantly affects the aorta, its major branches, and the pulmonary arteries. Segmental stenosis, occlusion, dilatation, or aneurysm formation may occur in the vessel wall during the course of the disease. Various signs and symptoms such as constitutional features (fever, malaise, anorexia, and weight loss), extremity pain, claudication, lightheadedness, bruises, absent or diminished pulses and reduced blood pressure can be present according to the vessel involved. Four cases of Takayasu arteritis were reported in the literature. These patients were females with age ranging from 29 to 68 years. Three of them had diffuse skin thickening. Computed tomographic angiography in all cases showed stenosis of various aortic branches. But peripheral gangrene is rare and only a few case reports are available.

Very few cases of scleroderma overlap syndrome with Takayasu arteritis were reported in the literature. Four cases of Takayasu arteritis in the setting of SSC have been reported. These patients were females with age ranging from 29 to 68 years. Three of them had diffuse skin thickening. Computed tomographic angiography in all cases showed stenosis of various aortic branches. Our case highlights the importance of being aware of a possible overlap syndrome when clinical features do not fit into one rheumatological disorder.

References