Summary

A 32-year-old female with a 10 year history of progressive skin thickening of her face, hands and feet along with raynaud's phenomenon and polyarthritis, presented with painful swelling and black discolouration of all her toes over 2 months. She also complained of fever and dysphagia over one month. Previously she had recurrent digital ulcers with secondary raynaud's and arterial duplex studies revealed reduced perfusion of her lower limbs. Prior to this admission she had undergone cardiac and pulmonary function tests which showed mild pulmonary hypertension and severe restrictive lung disease respectively. At that time she was tested positive for Anti-nuclear antibodies and Anti-U1 RNP antibodies hence suspected of mixed connective tissue disorder with scleroderma features. Therefore she was on methotrexate once weekly, prednisolone, nifedipine and sildenafil. On general examination there was tight thickening of the skin involving her face, hands and feet. Her toes on both feet were necrosed consistent with dry gangrene and there were discharging wounds proximal to the toe gangrene. On cardiovascular examination her left radial and bilateral dorsalis pedis pulses were absent whilst her other peripheral pulses were present. She had cold extremities with a prolonged capillary refilling time of 4 seconds. Precordial examination revealed a sustained left parasternal heave, a loud pulmonic component of the second heart sound and pansystolic murmur over the tricuspid area in keeping with tricuspid regurgitation. On chest examination there was dull percussion note in the lower lung zones bilaterally and bibasal fine end inspiratory crepitations. Her blood investigations showed elevated inflammatory markers with a neutrophil leucocytosis. Then she underwent a lower limb angiogram which showed proximally narrowed bilateral anterior tibial arteries whereas the distal anterior tibial arteries and right posterior tibial artery had absent flow. In the light of these findings she was reviewed by the vascular surgeon who opted for medical management. Therefore she received intravenous clindamycin and intravenous ceftazidime for one week along with daily washing of infected toe gangrene with hypertonic saline. She also received intravenous epoprostenol infusion daily for 5 days following which the patient symptomatically improved. Therefore she was discharged with oral antibiotics and followed up at rheumatology clinic a week later by which time her toe gangrene had worsened. We have diagnosed her with diffuse cutaneous systemic sclerosis complicated with pulmonary hypertension and bilateral toe gangrene.