Large vessel disease as a potentially treatable cause of devastating critical digital ischaemia in systemic sclerosis

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A 60-year-old lady with known limited cutaneous SSc (anti-centromere antibody positive) and a history of severe digital vascular disease presented with progressive digital vascular ischaemia. Her history included bilateral (periarterial) sympathectomy and digital amputation. She had no modifiable cardiovascular risk factors and was taking sildenafil for SSc-digital vasculopathy.

At presentation, there was ulceration of tip of the left index finger and thumb, with progressive necrosis (a) (Fig. 1). She was in significant pain and received a 5-day course of intravenous iloprost. Despite this, she developed very tender swelling of her left wrist and forearm with absent radial and brachial pulses.

A more proximal cause for compromised flow was suspected, and therefore, an urgently requested arterial Duplex scan demonstrated an occluded radial artery, with reversed flow in the ulnar artery. Flow in the brachial artery was noted to be severely damped. Therefore, computerised

tomography angiography (CTA) was performed with a view to therapeutic intervention.

CTA demonstrated a short occlusion of the left subclavian artery (b), which was confirmed by a catheter angiogram (c), performed prior to placement of a stent (d) to successfully relieve the obstruction. Subsequently, her clinical condition improved with reduced pain and limitation of ischaemic tissue loss.

Critical digital ischaemia in SSc is a medical emergency because early recognition of proximal (large) vessel disease can potentially be amenable to tissue-saving revascularisation [1–3]. An increased risk of macrovascular disease has been reported in SSc, in particular, in those with limited disease and positive anti-centromere antibodies (like our patient) [1–3]. Other potential causes include (but are not limited to) inflammation (e.g. vasculitis), infection and embolism [3]. Clinicians must maintain a high index of suspicion and always actively consider large vessel disease in SSc-related critical digital ischaemia.

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Fig. 1 Ulceration and necrosis of the left index finger and thumb (a), occlusion of the left subclavian artery on CTA (b) and catheter angiogram (c) which was stented (d)

Compliance with ethical standards

Disclosures None.

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