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Systemic sclerosis presenting with severe digital ischemia. A rare case report.

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Abstract: *Digital ischemic loss is a cause of significant morbidity in patients with systemic sclerosis (SSc). Both the small and large digital arteries are involved causing perfusion defects leading to ischemia. Microvascular disease causes intimal proliferation and luminal narrowing of small digital arteries, macrovascular disease causes narrowing or occlusion of larger digital arteries. Immediate clinical evaluation and treatment are mandatory at the onset of critical digital ischemia to prevent digital loss. We present a case of 38 year old female suffering from systemic sclerosis who presented with acute onset severe digital ischemia of all four limbs.*

Key words: *systemic sclerosis, ischemia, micro vascular, macrovascular*

I. Introduction

Systemic sclerosis (SSc) is a disease of unknown etiology characterized by immune activation, tissue fibrosis, and vasculopathy. Peripheral vascular involvement manifesting as Raynaud's phenomenon (RP) affects almost all patients. Episodes of progressive digital ischemia can result in a digital ulcer and sustained reduction in digital perfusion with impaired tissue viability can lead to critical ischemia, in some cases resulting in gangrene necessitating amputation. Neuroendothelial imbalance of vasoconstriction and vasodilatation, structural abnormalities of the vasculature, and intravascular factors such as platelet activation, procoagulants, and oxidative stress are the predominant factors that affect the microvasculature and cause critical ischemia. Recent studies on macrovascular disease in SSc suggest that ischemic demarcation and loss of digits occurs secondary to narrowing or occlusion of larger digital arteries (vessels of the palmar arch, radial, or ulnar artery) or medium-sized and large arteries in the lower extremities.

II. Case Report

A 38 year old female , house wife, diagnosed as limited systemic sclerosis 2 years back presented with gradually progressive painful discolouration of digits of the upper and lower extremities since 7 days duration. There was history of presence of Raynaud's phenomena since 3 years.

Her treatment history included tab. Prednisolone 10 mg alternate days, tab. Nicardipine 10 mg once daily, and tab. Pentoxiphylline 400mg 8 hourly.

On examination there was obvious bluish black discolouration of all the digits of the upper limbs including the hands and lower limbs upto the ankle joint bilaterally with visible line of demarcation. (Figure 1,2) Skin of the extremities (sclerodactyly) and face were thickened. Bilateral hands and feet were cold. Bilateral radial arteries were palpable, but in the lower limbs bilateral dorsalis pedis and posterior tibial arteries were not palpable. Allen's test was abnormal with poor refill bilaterally. Capillaroscopic examination of the periungal regions revealed dilated capillary loops. No peripheral bruits were audible.

An angiogram revealed evidence of a bilateral obliterative vasculopathic process in the superficial and deep palmar arches in the upper limbs and thrombotic occlusion of bilateral popliteal arteries of the lower limbs. Radiographs of the hands did not reveal any bony abnormality. Further investigations revealed a positive antinuclear antibody with titer > 1260 and anticentromere specificity. ACA were confirmed by enzyme-linked immunosorbent assay (ELISA) at greater than 110 U/mL. Anti-double stranded DNA, antineutrophil cytoplasmic antibodies, anticardiolipin antibodies, C-reactive protein, complete blood count, and urinalysis were all normal or negative. Chest radiograph, echocardiogram were normal.

She was treated with intravenous heparin, IV methyl prednisolone, clopidogrel , pentoxifylline, intravascular thrombolysis with urokinase for the lower limb thrombotic disease. But there was no clinical improvement and the pain and necrosis increased further. The patient was referred to higher centre.

III. Discussion

SSc has a prevalence of 1–50 cases per 100,000 people worldwide. [1] A retrospective review of the clinical status of 98 patients with SSc, seen between 1985 and 1990, showed that amputation of 1 or more digits due to ischemia occurred in 20.4% of the patients while 9.2% had multiple digit loss as in our case. [2] Out of two prospective cohort studies in UK , one study found that 28 (16%) of 171 SSc patients had at least one digital amputation and 73 (43%) had experienced at least one episode of severe digital ischemia, [3, and in another study 17.4% were found to have complications related to severe digital vasculopathy including digital ulcers, critical digital ischemia, gangrene, or the need for digital sympathectomy. Furthermore, 1.6% of the total cohort developed critical digital ischemia, 1.4% developed digital gangrene, and 0.9% required amputation. [4]
Observational studies have shown that limited cutaneous SSc is associated with more prominent vasculopathy than its diffuse counterpart.

Several studies have demonstrated that the severity of the vasculopathy requiring amputation is associated with presence of specific antibodies like anti- cardiolipin , anti-centromere, anti-beta2-glycoprotein I , and anti-topoisomerase antibodies. [5, 6]
Smoking is also independently associated with severe digital gangrene. [7]

Evaluation should ideally start with meticulous clinical examination, palpation of peripheral pulses and assessing for persistent discoloration (cyanosis or pallor), digital ulceration, extreme tenderness, or frank gangrene. Nailfold capillaroscopic changes may be predictive of the development of digital ischemia. [8] Arterial Doppler, and Ankle brachial pressure index estimation is also helpful.

Laboratory analysis for prothrombotic states including the antiphospholipid antibodies (lupus anticoagulant, anti-cardiolipin, and anti-beta2-glycoprotein I antibodies), anti centromere antibodyshould be performed in all patients. Prompt clinical evaluation and referral for treatment is critical to the prevention of progression to digital loss.

Angiographic evaluation for digital occlusions include conventional angiography, magnetic resonance angiography (MRA) or computed tomography (CT) angiography. Conventional angiography is extremely sensitive for identifying stenosis, occlusion, aneurysm, or other vascular irregularities and is still considered gold standard.

IV. Treatment

Digit-threatening ischemia is a medical emergency. It warrants prompt and aggressive treatment to control symptoms and prevent digital loss.

Nonpharmacologic treatments aims towards keeping the limb warm and giving rest to the part with decreased activity. Xeroform dressing with an antibiotic ointment can be used to prevent a superinfection and allow wound healing. Intravenous antibiotics should be used to overlying infection or osteomyelitis. Surgical debridement is generally reserved for patients with purulent drainage, necrotic/late stage ischemic tissue, or severe structural arterial disease who do not respond to medical therapies.

Analgesia with opioids is of utmost importance. Local anesthetic blocks with lidocaine or bupivicaine without epinephrine may be helpful for pain control.
Anticoagulation with intravenous heparin is recommended.

Aggressive vasodilatation in form oral calcium channel blocker, intravenous prostacyclins, are considered the mainstay of management for acute digital ischemia. Intravenous epoprostenol or iloprost (0.5–2 ng/kg/min) daily infusions for 1–3 days, each infusion lasting 6 hours, is the recommended regimen. [9] If symptoms are persistent and medical therapy fails, proximal or digital sympathectomy, microsurgical revascularization of the hand, and digital arterial reconstruction have been reported to improve digital vascular perfusion, heal digital ulcers, and substantially relieve or eliminate pain. [10]

If there is larger vessel occlusive disease, especially at the level of the ulnar or radial artery, successful reconstruction can be performed with vein grafts. [11] Peripheral artery bypass is a good alternative in vasoocclusive disease of the superficial palmer arch and tarsal arch, and also in relieving symptoms in lower limb larger arterial occlusive disease.

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