

# Acute Presentation of Critical Limb Ischemia In A Patient With Scleroderma

Omer Nasim<sup>1\*</sup>

## ABSTRACT

*Systemic sclerosis is a disease of the connective tissue of unknown etiology. It is characterized by multiorgan involvement and a wide range of clinical manifestations. It rarely presented with acute critical limb ischemia. Here we report a case of 61-year old male who presented with severe pain in the lower right extremity. He also had mild cough and shortness of breath. It was later discovered through past medical history and previous investigations that he was a case scleroderma and its presentation was with acute limb thrombus for which amputation was performed.*

*Key words* Scleroderma, Amputation, Gangrene.

## INTRODUCTION:

The prevalence of systemic sclerosis in the different geographical areas ranges from 233-277 cases per million whereas the incidence of systemic sclerosis varies from 19-23 cases per million each year in different studies.<sup>1</sup> Survival rate of systemic sclerosis is low with multi-organ system involvement. A study showed the survival rate of patients with several organ involvement as 38% compared with no major organ involvement as 72%.<sup>2</sup> Leading cause of death in systemic sclerosis is pulmonary fibrosis followed by pulmonary arterial hypertension.<sup>3</sup> Clinical features of systemic sclerosis include skin change, pulmonary fibrosis, fibroproliferative vasculopathy and Raynaud's phenomenon.<sup>4</sup> Systemic sclerosis is classified into two forms: Diffuse cutaneous and limited cutaneous.

## CASE REPORT:

A 61-year-old male admitted to with shortness of breath and dry cough with failure to maintain oxygen saturation. High resolution chest CT scan showed patchy ground glass haze in lower lobes of both the lungs. The overall findings were consistent with idiopathic pulmonary fibrosis. The scan also showed hypodense foci in upper pole of the spleen. The patient was prescribed medicines and sent home. One month later he presented with severe pain in the lower right extremity. On examination, the lower

right extremity was pulseless, cold and cyanosed. On CT angiogram scan multiple floating atheromas were seen in the thoracic aorta and the right lower limb showed severe focal stenosis at the origin of superficial femoral artery, occluded popliteal artery with distal reconstitution and severe diffuse disease in distal anterior tibial artery and dorsalis pedis artery. The tibioperoneal trunk, proximal posterior tibial artery, distal anterior tibial artery and dorsalis pedis artery of the left lower extremity were also occluded on the CT scan. An emergency embolectomy in the lower right extremity was performed by a vascular surgeon after a diagnosis of lower limb ischemia.

In postoperative period after 48-hours patient complaint of severe abdominal pain and an urgent CT angiogram of mesenteric vessels revealed occlusion of distal segment of superior mesenteric artery (SMA) and gangrenous small bowel. The splenic artery was also occluded, and a large spleen infarct was observed. On emergency exploratory laparotomy, gangrenous distal ileum, cecum and ascending colon were seen and resected. No intestinal perforation was found. Histopathology reports of the ileum and ascending colon reported extensive hemorrhagic and gangrenous areas. Microscopic examination revealed severe acute on chronic inflammation, thrombosis and infraction. The appendix was unremarkable.

The patient was consulted by a pulmonologist during postoperative period when shortness of breath and dry cough were observed. Systemic sclerosis was suspected when history of pulmonary fibrosis was considered, and examination revealed sclerodactyly and digital telangiectasia. Autoimmune investigations were positive for ANA and Scl 70 antibody.

<sup>1</sup> Rehman Medical Institute Peshawar

## Correspondence:

Dr. Omer Nasim<sup>1\*</sup>

Rehman Medical Institute

Peshawar

Email: omer.nasim-12@rmi.edu.pk

During his stay at the hospital patient was started on heparin and then switched to warfarin. At the time of discharge his INR was 2.90 and he was prescribed warfarin, cephadrine, fluticasone and salbutamol. A month later he presented again with right lower limb pain and underwent a myectomy procedure followed by right lower limb amputation.

#### DISCUSSION:

The extent of vasculopathies play a crucial role in determining the prognosis of patients with scleroderma. Usually attention is focused on small vessel involvement. Microvascular disease is a well-known feature of scleroderma which commonly manifests as Raynaud's phenomenon. Increased risk of ischemic digital loss has been reported in other studies.<sup>5</sup> Pathological studies on changes in small vessels show concentric intimal proliferation, medial thickening and fibrous adventitial thickening.<sup>6</sup> Macrovascular disease in scleroderma has largely been unrecognized and only a small number of case reports on macrovascular involvement are found. All of these cases were associated with significant mortality and morbidity.<sup>7</sup>

The patient presented in this report had no signs of microvascular complications that are usually present in a typical scleroderma case. Lack of history of microvasculature abnormalities such as Raynaud's phenomenon or digital ulcers can be attributed to his delayed diagnosis of scleroderma. The only signs of scleroderma present initially were sclerodactyly and pulmonary fibrosis but the urgent nature of this case in our hospital lead to a late diagnosis. Systemic scleroderma was only suspected when the involvement of macro-vasculature along with sclerodactyly and history of pulmonary fibrosis was considered.

Patients with severe scleroderma are predisposed to developing leg ulcers and gangrene. Our patient presented with severe intermittent claudication in the right leg and was found to have stenosis in the superficial femoral artery and occluded popliteal artery. Peripheral arterial disease and venous insufficiency was found in 50% of patients in a study done on legs ulcers in systemic sclerosis.<sup>8</sup> Hasegawa et al reported involvement of ulnar and radial artery in 42% of their patients and only one patient had occluded posterior tibial artery and dorsalis pedis artery.<sup>9</sup> In our case there was no evidence of signs of upper limb occlusion or ischemia however the diffuse disease was seen in tibial artery and dorsalis pedis artery on imaging. Our patient one month after an embolectomy in the right lower limb presented with muscle necrosis for which he went under

myectomy followed by the right lower limb amputation. Our patient had positive titers of the anti-scl-70 antibody and was an undiagnosed case of scleroderma. Patients with systemic sclerosis who present with either clinical signs or radiographic evidence of pulmonary fibrosis but no evidence of cutaneous involvement can later develop scleroderma. An esophagogram and antinuclear antibody test may help facilitate diagnosis of this condition.

#### REFERENCES:

1. Nikpour M, Stevens WM, Herrick AL, Proudman SM. Epidemiology of systemic sclerosis. *Best Pract Res Clin Rheumatol.* 2010;24:857-69. <http://dx.doi.org/10.1016/j.berh.2010.10.007>
2. Steen VD, Medsger TA. Severe organ involvement in systemic sclerosis with diffuse scleroderma. *Arthritis Rheum.* 2000;43:2437-44.
3. Tyndall AJ, Bannert B, Vonk M, Airò P, Cozzi F, Carreira PE, et al. Causes and risk factors for death in systemic sclerosis: a study from the EULAR Scleroderma Trials and Research (EUSTAR) database. *Ann Rheum Dis.* 2010;69:1809-15.
4. Masi AT, Rodnan GP, Medsger Jr. TA, Altman RD, D'Angelo WA, Fries JF, et al. Preliminary criteria for the classification of systemic sclerosis (scleroderma). Subcommittee for scleroderma criteria of the American Rheumatism Association Diagnostic and Therapeutic Criteria Committee. *Arthritis Rheum.* 1980;23:581-90.
5. Wigley FM, Wise RA, Miller R, Needleman BW, Spence RJ. Anticentromere antibody as a predictor of digital ischemic loss in patients with systemic sclerosis. *J Am Coll Rheumatol.* 1992;35:688-93.
6. Campbell PM, LeRoy EC. Pathogenesis of systemic sclerosis: a vascular hypothesis. *Semin Arthritis Rheum.* 1975;4:351-68. doi:10.1016/0049-0172(75)90017-7
7. Barile LA, Bravo G, Pizarro S. Large vessel arterial thrombosis in systemic sclerosis with antiphospholipid antibodies. *J Rheumatol.* 1990;17:1721.

8. Hafner J, Schneider E, Burg G, Cassina PC. Management of leg ulcers in patients with rheumatoid arthritis or systemic sclerosis: The importance of concomitant arterial and venous disease. *J Vasc Surg.* 2000;32: 322-9.

Received for publication: 24-08-2019

Accepted after revision: 30-03-2020

Author's Contributions:

Omer Nasim: Data collection and manuscript writing.

9. Hasegawa M, Nagai Y, Tamura A, Ishikawa O. Arteriographic evaluation of vascular changes of the extremities in patients with systemic sclerosis. *Br J Dermatol.* 2006; 155: 1159-64. <https://doi.org/10.1111/j.1365-2133.2006.07475.x>

Conflict of Interest:

The authors declare that they have no conflict of interest.

Source of Funding: None

How to cite this article:

Nasim O. Acute presentation of critical limb ischemia in a patient with scleroderma. *J Surg Pakistan.* 2019;25(2):96-98.

Doi:10.21699/jsp.25.2.11.