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Case report Open Access

Bilateral popliteal artery occlusion in a young woman as a rare complication of systemic sclerosis: A case report

Mohammad Hassan Jokar[1], Nima Zafari[2], Mahla Velayati[3], Kamila Hashemzadeh[4]*

[1] Assistant professor of community medicine, Department of community medicine, School of Medicine, Rafsanjan University of Medical Sciences, Rafsanjan, Iran. [2] Assistant Professor of Psychiatry, Department of Psychiatry, School of Medicine,Moradi Educational and Treatment Hospital, Rafsanjan University of Medical Sciences, Rafsanjan, Iran. [3] General Physician, Clinical Research Development Unit, Ali-Ibn Abi-Talib Hospital, Rafsanjan University of Medical Sciences, Rafsanjan, Iran. [4] Department of Internal Medicine, School of Medicine; Clinical Research Development Unit, Ali-IbnAbi-Talib Hospital, Rafsanjan University of Medical Sciences, Rafsanjan, Iran.

Systemic sclerosis (SSc) is an autoimmune rheumatic disorder characterized by vascular damage, sclerotic skin changes, and multi-system involvement of internal organs. Digital ischemic lesions, nailfold capillary abnormalities, and secondary Raynaud's phenomenon are common manifestations of microvascular injuries. Recent findings have revealed that macrovascular involvement in SSc more prevalent than previously believed. There have been reports of large vessel involvement, such as the ulnar, femoral, carotid, renal, and pulmonary arteries. We report here a case of a young woman with progressive SSc who complained of lower limb pain, dysphagia secondary to esophageal dysfunction, and urinary incontinence.She had symptoms of Raynaud's phenomenon, skin sclerosis, sclerodactyly, digital pitting ulcers,nailfoldcapillary abnormalities, and digital ischemia. On special examination, the anti-centromere antibody was detected. She developed bilateral popliteal artery occlusion as a complication of SSc.

Keywords: Systemic sclerosis; Scleroderma; Macrovascular involvement; Popliteal artery occlusion; Case report

Introduction

Systemic sclerosis (SSc) is an immune-mediated rheumatic disease with unknown etiology, characterized by vascular damage, sclerotic changes in the skin, and multi-system involvement of internal organs (kidney, lung, gastrointestinal tract, and cardiovascular system). Common manifestations of SSc such as digital ischemic lesions, nailfold capillary abnormalities, and secondary Raynaud's phenomenon reflect the widespread systemic microvascular injuries [1]. Previous studies have suggested that macro-vasculopathy is considered occasional in SSc [2]. However, recent findings have revealed that macrovascular changes inthe peripheral artery are more prevalent than previously believed. There have been reports of large vessel involvement such as the ulnar, femoral, carotid, renal, and pulmonary arteries 3-5. We present a young woman with SSc who developed bilateral popliteal artery occlusion

Case pescription

In March 2003, a 32-year-old woman was admitted to our rheumatology clinic. She complained of dysphagia secondary to esophageal dysfunction,

*Corresponding Author: Kamila Hashemzadeh, Rheumatic Diseases Research Center, Mashhad University of Medical Sciences, Mashhad, Iran. Email: hashemzadehk@mums.ac.ir
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chest pain, right lower limb pain, and urinary incontinence. Her clinical manifestations included skin sclerosis, sclerodactyly, the Raynaud phenomenon, digital pitting ulcers, and digital ischemia. There was no history of risky habits such as smoking and other risk factors for vascular disease such as diabetes mellitus or dyslipidemia. Blood pressure was controlled near the normal range with a calcium channel blocker. The anti-centromere antibody was detected on special examinations. Laboratory data showed normal cell blood count, hemoglobin: 10.3 g/dL (normal range for women: 11.0 –15.5 g/dL) indicating mild anemia, hematocrit: %34, erythrocyte sedimentation rate: 25 mm/hr. C-reactive protein and serum total cholesterol were within the normal range. Coagulation tests and liver function tests were normal. Echocardiography showed mild mitral regurgitation and tricuspid regurgitation without evidence of vegetation or intracardiac thrombus. A High-resolution computed tomography (HRCT) scan of the chest showed no evidence of interstitial lung disease (ILD). Based on clinical observation and serological assessments, she was diagnosed with progressive systemic sclerosis (PSS) and hospitalized for further management.

Medications included acetylsalicylic acid for digital ischemia, diltiazem (with ahistory of previous intolerance of nifedipine), omeprazole for dysphagia, prednisolone, warfarin, and folic acid. Additionally, the patient received cyclophosphamide and azathioprine for her major skin involvement. Moreover,the patient was depressed, referred to a psychiatrist, and treated with fluoxetine. In August 2013, the patient developed cellulitis in the right forearm and elbow which was treated by cefalexin. Although warfarin was administered to improve peripheral circulation, the right lower limb pain and toe necrosis were considerably worsening.

After February 2015, our findings revealed subcutaneous calcium deposition in the upper part of the right thigh and left shoulder (calcinosis cutis), inflammatory ulcers of the fingers, toes, and heels. During the next 7 months, the patient demonstrated chest pain and dyspnea. She was referred to a cardiologist and was diagnosed with ischemic heart disease (IHD). Despite previous treatments, in May 2020, the patient came to our clinic complaining of lower limb pain exacerbation, ankle swelling, and aright wrist infection. On physical examination, the popliteal pulse was weak. Intra-arterial CT angiography (Figure1) and three-dimensional CT angiography (Figure2) of the lower limb revealed occlusion of the popliteal arteries and significant narrowing in the peroneal and tibialis (anterior and posterior) arteries. Femoral, internal, and external iliac arteries were normal.

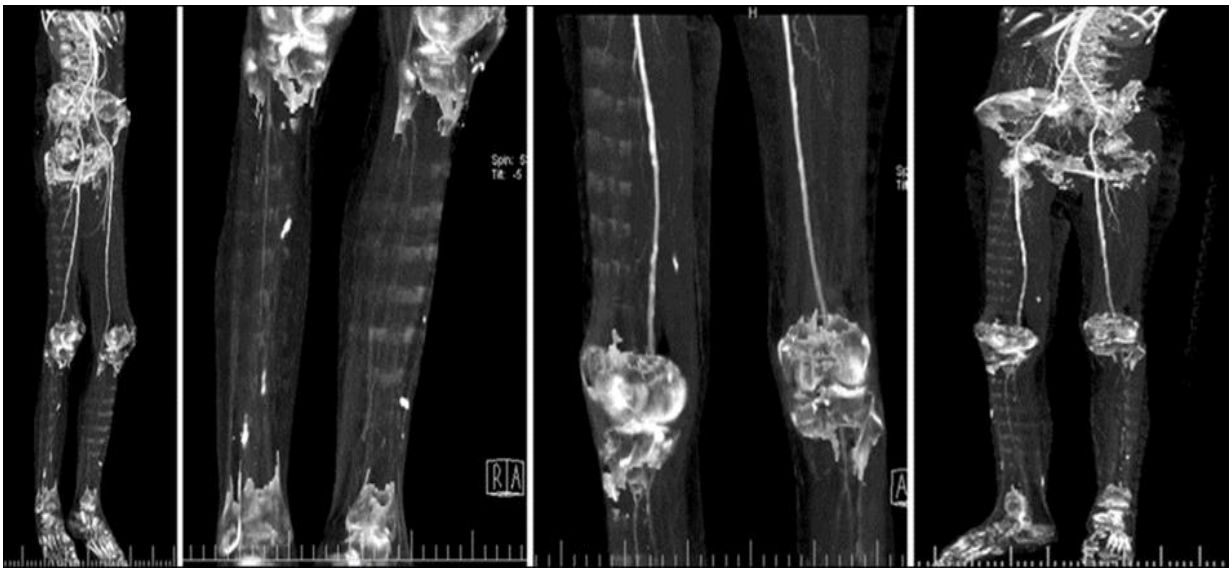

Figure 1. The angiography demonstrated subtotal occlusion at the proximal popliteal artery near to plateau of the tibia, and narrowing in the peroneal anterior, and posterior tibialis arteries.

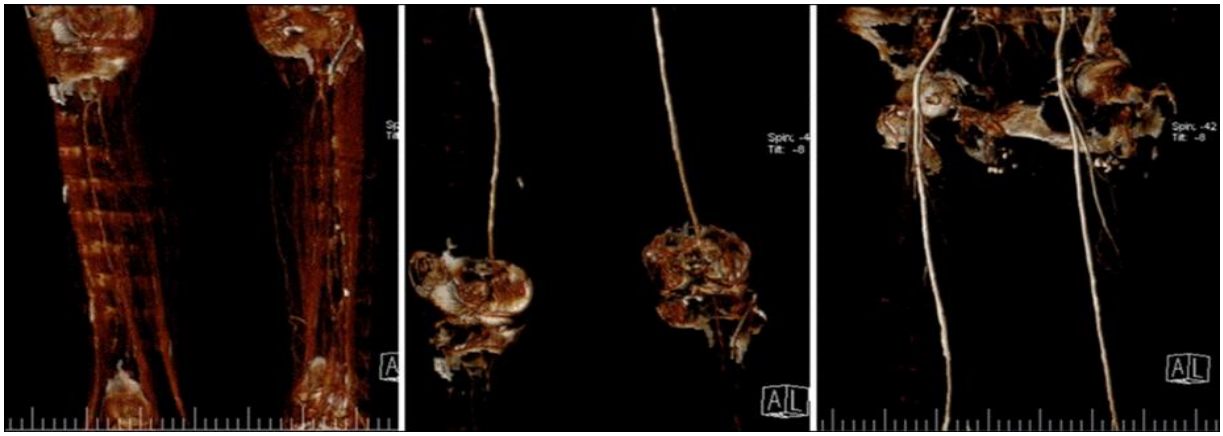

Figure 2. Three-dimensional computed tomography angiography shows popliteal occlusion.

SSc is a connective tissue disorder characterized by skin and internal organs fibrosis and vasculopathy. Vascular involvement is a well established component of SSc and a critical factor for the prognosis. Vasculopathy in SSC can lead to clinical manifestations, including Raynaud's phenomenon, digital ulcers, gangrene, pulmonary arterial hypertension, and scleroderma renal crisis [6]. Recent studiesc have demonstrated that macroangiopathy can accompany SSc, although its frequency is lower than peripheral micro-vasculopathy, such as Raynaud'sphenomenon [7]. Large vessel involvement characterized by medial thickening with circumferential luminal narrowing and occlusion with an acellular material can be due to the pathophysiology of systemic sclerosis and concomitant atherosclerosis. Inflammation, persistent vasospasm of small vessels, endothelial dysfunction, increased levels of lipoprotein A, and oxidized low-density lipoproteins (LDL) associated with scleroderma can contribute to macroangiopathy, as well as additional risk factors such as smoking, age, diabetes mellitus, or dyslipidemia [8]. Recent observations have shown that macrovascular disease and small vessel injuries may lead to digital ulcers and gangrene in patients with systemic sclerosis. Cases of SSc with macro-vasculopathy include the involvement of the subclavian artery, common carotid artery, ulnar, femoral, popliteal, mesenteric, anterior, and posterior tibial arteries [3,5]. In 1980, Borra SI et al. reported a 42-year-old woman with renal failure due to progressive SSc in whom renal angiography showed bilateral renal artery occlusion [4].

In 2011, YoheiIchimura et al. described a case of SSc in a 50-year-old Japanese woman with ulnar artery stenosis, which was evaluated by magnetic resonance angiography and diminished by the bosentan treatment [2]. In 2017, Rin Shimozato et al. reported a 75-year-old woman with carotid artery stenosis suspected to have been induced by SSc who was treated by carotid artery stenting (CAS) [9]. Our patient is a further case of clinical macro-vessel involvement in SSc. There have been case reports linking the presence of anti-centromere antibodies to large and medium vessel involvement. Anticentromere antibodies, smoking and drinking, older age, dyslipidemia, diabetes mellitus, and obesity are risk factors for developing macroangiopathy in SSc [9]. Anti-cardiolipin antibody (ACL) is another factor affecting large vessels and involves thrombus formation. This antibody was detected in patients with systemic lupus erythematosus, idiopathic thrombocytopenic purpura, anti-phospholipid antibody syndrome, and progressive SSc [10]. However, this antibody was negative in the case we described here.

This woman had no clinical evidence of significant underlying atherosclerosis and traditional risk factors for peripheral vascular disease. Previous reports suggest that it is necessary to consider atherosclerosis as an underlying cause in the presence of risk factors such as diabetes, hyperlipidemia, or unilateral vascular lesions. Despite the low-risk status of macrovasculopathy, our patient developed bilateral popliteal artery stenosis, so the role of scleroderma was considered highly probable. Despite the low-risk state of macrovasculopathy, our patient developed bilateral

popliteal artery stenosis, so the large vessel involvement in SSc was considered most likely.

Conclusion

Here we present a patient with SSc with bilateral obstruction of the popliteal arteries. Although SSC is a rare cause of large peripheral vascular disease, clinicians should pay special attention to the history and physical examination of patients with SSc complaining of pain, digital pitting ulcers, digital ischemia, and other evidence of large vessel involvement. A high level of clinical suspicion is necessary for early diagnosis and preventing complications of macrovascular involvement, such as gangrene and subsequent amputation.

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Conflicting Interests

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