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Case Report

A case of serpentine coronaries and acute myocardial infarction

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ABSTRACT

Microvascular disease is a prominent feature of systemic sclerosis (SSc) and leads to Raynaud's phenomenon, pulmonary arterial hypertension, and scleroderma renal crisis. The presence of macrovascular disease is less well established, and, in particular, it is not known whether the prevalence of coronary heart disease in SSc is increased. We report a case of SSc who presented with evolved myocardial infarction whose angiogram revealed tortuous coronaries and peripheral arteries. Regional wall motion abnormality was not demonstrated on echocardiography. The microvascular dysfunction and vasospasm of coronaries were responsible for the myocardial infarction.

Keywords: Tortuous coronaries, Systemic sclerosis, Microvascular dysfunction

INTRODUCTION

The literally scleroderma means hard skin. But this disorder is more than just skin involvement as it can affect multisystem in the human body. Systemic sclerosis (SSc) is a chronic autoimmune disease which can affect the skin and various internal organs.[1] Apart from scleroderma it is characterised by vasculopathy, Raynaud's phenomenon and auto anti bodies. Acute myocardial infarction is an uncommon (1%) manifestation in systemic sclerosis patients. Normal coronaries are seen more commonly in these patients as compared to the general population.[2] Below we present a case of systemic scleroderma who presented with myocardial infarction.

CASE REPORT

A 64 year lady, an Agriculturist presented with head ache and vomiting of 2 days duration. She had typical chest pain suggestive of angina with dyspnea of one day duration. This patient had significant past medical history. She had multiple large and small joint pain for the last five years. Intermittently she was having swelling of fingers in both the hands. She also complained of ulcers in the finger tips for the last one year. Swelling and pain used to increases on exposure to cold which was suggestive of Reynaud's phenomenon. She had significant loss of weight in the past 3 months with loss of appetite. She also had difficulty in swallowing since one month. There was no history of dryness of mouth and eyes. Recently she had developed non productive cough. For the past one year she was treated by local doctors with intermittent steroids for arthritis. This patient was a first of six siblings, all other siblings were healthy.

On examination she was hemodynamically stable, her palpebral conjunctiva was pale. Digital pitting ulcers were present at the tip of the fingers (see fig 1 (a)). Sclerodactyly and thickening of skin in the distal extremity and face was noted. She had decreased eversion of eye lids. She had positive Raynaud's phenomenon. Cardiovascular examination showed normal cardiac sounds with no added sounds, respiratory system examination showed end expiratory fine

crepitations. There was no organomegaly on per-abdomen examination.

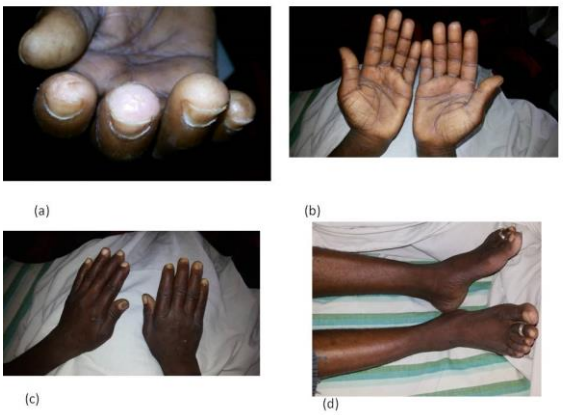

Figure 1: Photographs of hands and feet, showing thickening of skin and digital pitting ulcers in both hands.

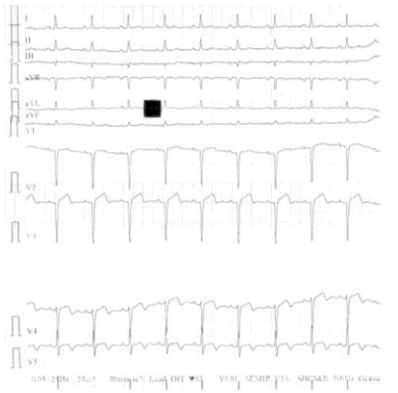

Figure 2: ECG showing evolved Anterior wall MI.

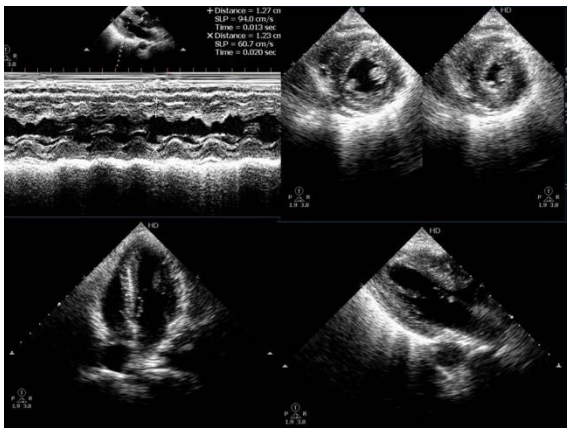

Figure 3: 2D-Echocardiograph showing normal chambers with no regional wall motion abnormalities.

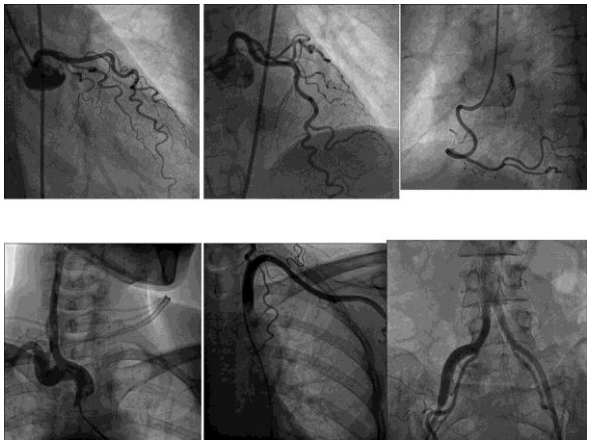

Figure 4: Angiogram showing tortuous coronaries, and proximal peripheral arteries.

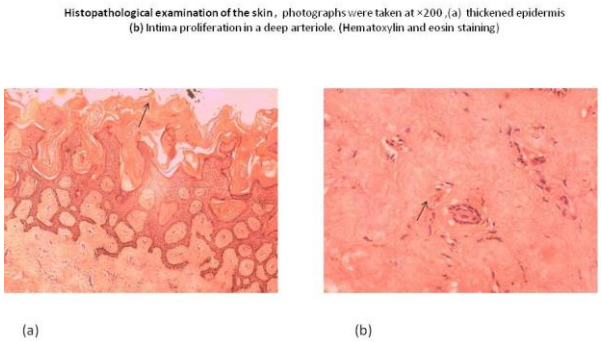

Figure 5: Skin biopsy showing thickened epidermis, with obliteration of vascular lumen in dermis.

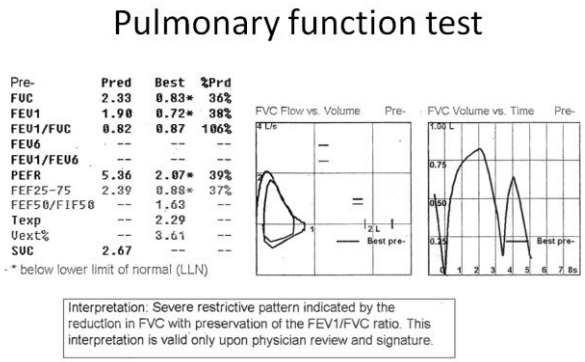

Figure 6: Pulmonary function tests shows restrictive pulmonary disease pattern.

On investigation revealed a positive troponin–T (0.208 ng/ml [normal<0.01ng/ml]), CPK was 386 U/L, CKMB of 46 U/ml. ESR was 8mm /hour, Lipid profile, renal function and liver function tests were within normal limits. ANA was strongly positive (1:40 (4+) nucleolar type), SCL-70 was detected in the serum suggestive of diffuse Scleroderma. ECG showed changes of evolved anterior wall myocardial infarction. Chest x ray was

normal. Pulmonary function tests revealed restrictive lung disease. Echo cardiograph showed no regional wall motion abnormalities and had adequate left ventricular function. Her coronary angiogram revealed tortuous coronaries no obstruction, even peripheral angiogram showed tortuous vessels. Skin biopsy revealed increased thickness of epidermis, with intimal proliferation and obliteration of lumen of arterioles.

DISCUSSION

In patients who did not had atherosclerotic coronary artery disease cardiac wall motion abnormalities on echocardiography was surprisingly absent, though concentric LVH was uniformly present. In contrast, in patients who had atherosclerotic disease, there were pronounced myocardial motion abnormalities without LVH. These observations suggest that coronary ischemia related to coronary vasospasm may be responsible for the occurrence of MI in patients with SSc.[2] SSc is independently associated with risk of AMI development.[3] Microvascular disease is a prominent feature of systemic sclerosis (SSc). The presence of macrovascular disease is less well established, and, in particular, it is not known whether the prevalence of coronary heart disease in SSc is increased.[4]

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