Systemic sclerosis, localized morphea, en coup de sabre and aortic regurgitation: A rare association

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ABSTRACT

A 24-year-old male presented with Raynaud's phenomenon, digital infarcts, salt and pepper pigmentation and characteristic facies. There was gastrointestinal involvement clinically, endoscopically as well radiologically. In view of these findings and the demonstration of anti-nuclear antibodies with a homogeneous pattern of immunofluorescence and ScL-70 antibodies, he was diagnosed as a case of diffuse cutaneous systemic sclerosis. In addition, the patient had plaques of localized morphea, en coup de sabre with facial hemiatrophy on the left side, and mild aortic regurgitation (detected on echocardiography). The occurrence of these rare associations of localized plaques of morphea, en coup de sabre and aortic regurgitation in a single case of systemic sclerosis is quite an exceptional and interesting occurrence.

Key Words: Systemic sclerosis, Localized morphea, En coup de sabre, Aortic regurgitation

INTRODUCTION

Localized morphea or scleroderma is a disorder of unknown cause in which there is localized sclerosis of the skin. It can have circumscribed plaques or bands, linear morphea or fronto-parietal lesions (en coup de sabre), with or without facial hemiatrophy. On the other hand, systemic sclerosis is quite distinct with endothelial changes in the muscle capillaries causing damage to various organs like skin, gastrointestinal tract, lungs, heart, kidneys, bones, muscles, etc.[1] The two disorders rarely occur together.[2] A few cases of systemic sclerosis with en coup de sabre have been reported.[3,4] Further, aortic regurgitation is rare in systemic sclerosis.[5-7] We describe here a patient of systemic sclerosis who had clustering of three rare associations viz. localized morphea, en coup de sabre and aortic regurgitation.

CASE REPORT

A 25-year-old male presented with history of Raynaud's phenomenon over fingers and toes since 3 years, recurrent painful ulcers over fingers and toes since 2½ years, tightening of the skin since 2 years, postprandial odynophagia to liquids since one year. There was no history of palpitations, dyspnea, syncope, cough or pain chest. The course was progressive and unremitting.

The patient's pulse, blood pressure and respiratory rate were normal. Chest expansion was restricted to only one cm at the level of the nipples and the underlying skin was sclerotic. The chest was clear, without any adventitious sounds. The cardiovascular and other

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systems were essentially normal.

The patient had a midline depression of the forehead and hemiatrophy involving the left half of the face with multiple hyperpigmented, sclerotic plaques having central ivory white depigmentation over the frontal, parietal and mastoid regions of the scalp, bridge of the nose and both aural helices. Similar plaques were also seen over the palate buccal mucosa bilaterally. A solitary patch of non-scarring alopecia involving the left half of the upper lip was present. His skin of the face and scalp was shiny, waxy, smooth, and inelastic and the facies mask like with a pinched nose, microstomia and positive Ingram's sign. In addition, there were multiple well defined and diffuse hyperpigmented, sclerotic plaques with salt and pepper pigmentation over the left supraclavicular region, shoulders, hypogastrium, and both iliac and trochanteric regions varying in size from a few to many centimeters. He also had acrosclerosis with semiflexed fingers, loss of fingertip pulp, multiple stellate scars and small ulcers over the fingertips, shiny curved nails, cyanosed fingers and toes, and ragged cuticles. There was also generalized hyperpigmentation of the skin.

Hemogram, urinanalysis, liver function tests, blood urea, serum creatinine, ultrasound abdomen, ECG and radiograph of the chest were normal. LE cell test and RA factor were negative and VDRL was non-reactive. The skin biopsy was consistent with the diagnosis of scleroderma. ANA test was positive with a homogeneous pattern on immunofluorescence. The

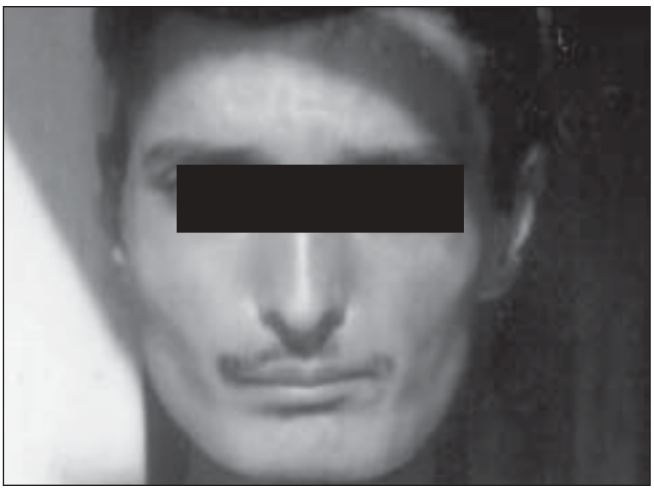

Figure 1: Mask-like facies, pinched nose, waxy forehead and sclerotic plaque over the left fronto-parietal region

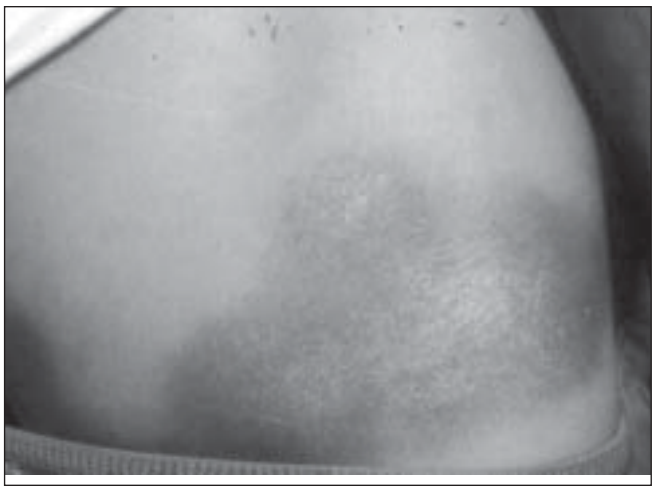

Figure 3: Sclerotic plaque over the right buttock

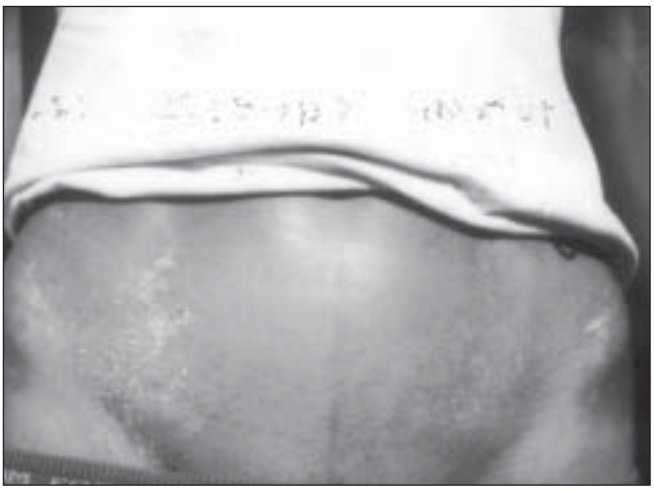

Figure 2: Sclerotic plaques with salt and pepper pigmentation over the iliac regions

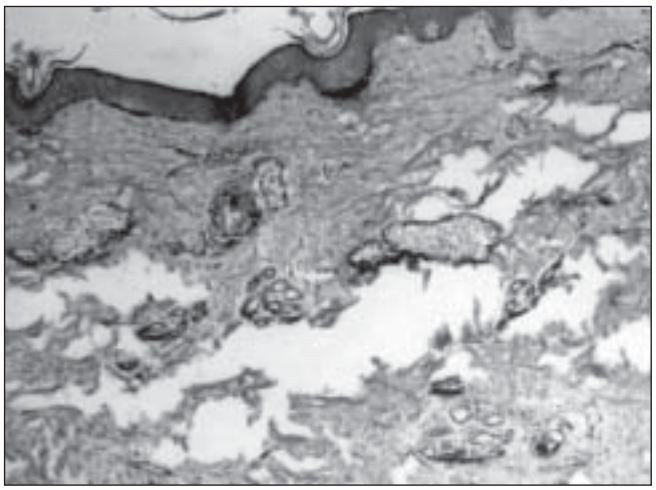

Figure 4: Increased collagenization with a mild mononuclear infiltrate in the dermis

test for ScL-70 antibody was positive while tests for anti-Ro, anti-La, anti-Sm, anti-RMP and anti-Jo1 antibodies were negative. The pulmonary function test showed moderately severe restrictive lung disease. Upper GI endoscopy revealed esophageal dilatation, dysmotility and reflux esophagitis. Barium meal follow-through showed dilatation of the lower part of the esophagus, stomach and second part of the duodenum. Mild aortic regurgitation was detected on echocardiography.

Diagnosed as a case of diffuse cutaneous systemic sclerosis with the associations of en coup de sabre, localized morphea and mild aortic regurgitation, the patient has been put on monthly intravenous pulses of dexamethasone 100 mg daily on three days, cyclophosphamide 500 mg on day one and a daily oral dose of 500 mg of cyclophosphamide. The patient has received twelve such pulses till date and is showing good improvement.

DISCUSSION

Due to the presence of sclerosis of the skin above the elbows, acrosclerosis, Raynaud's phenomenon, digital infarcts, salt and pepper pigmentation, characteristic facies, gastrointestinal involvement, positive ANA test with homogeneous pattern of immunofluorescence and ScL-70 antibodies, there was no doubt regarding the diagnosis of diffuse cutaneous systemic sclerosis. However, the presence of a plaque of localized morphea and en coup de sabre was quite interesting as only a few cases of association of systemic sclerosis with localized scleroderma,[2] and en coup de sabre[3,4] have been reported. Nomura et al[3] reported a female having en coup de sabre on the right side, along with symptoms of systemic sclerosis more on the same side. Aortic valve involvement causing aortic regurgitation is rare in cases of systemic sclerosis.[5-7] The presence of these rare associations in a single patient makes our case exceptionally interesting. Our case also illustrates that detailed investigations in a patient of systemic sclerosis to pick-up early involvement of various organs are amply justified as revealed by the detection of mild aortic regurgitation only on echocardiography.

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