Scleroderma with Rash and Clubbing – A Case Report

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Introduction

Skin in scleroderma can be affected with or without systemic involvement. Lesions have been described as localized, generalized, or linear morphoea¹. The patient described had clubbing and guttate depigmented plaques. Acquired clubbing has not been described previously, but guttate lesions have¹.

Case History

A 24 year old previously healthy, Jamaican-born man presented with a nine-month history of half a stone weight loss, general malaise, inability to pass the army basic fitness test due to shortness of breath, skin rash and Raynaud's phenomenon.

There was no history of previous illness, in particular there was no pulmonary disease, exposure to industrial material, infectious diseases or chronic diarrhoea. There was no history of clubbing in the family.

On examination, there was clubbing, multiple splinter haemorrhages in the proximal nail folds and taut, shiny distal digital skin (Fig 1). The upper part of the body was covered with multiple, one centimetre diameter depigmented plaques (Fig 2). There was no lymphadenopathy or hepatosplenomegaly, heart sounds were normal, and direct questioning elicited dysphagia.

Investigations

The following were abnormal:-

ESR 55 mm/1st hr, antinuclear factor (diffuse pattern titre 1 in 640). Endoscopy showed mild duodenitis with

Fig 1. Clubbing with early drum stick formation. Also showing splinter haemorrhages in the proximal nail fold and shiny distal digital skin.

Fig 2. Skin rash – depigmented spots.

no abnormal oesophageal peristalsis. Skin biopsy from an abdominal wall lesion revealed "sclerosis of dermal collagen with some focal areas of perivascular infiltration of lymphocytes in the dermis".

Vitalograph: Forced expiratory volume in first second 3.60 L, forced vital capacity 4.42 L, FEV₁/FVC 81.2%. Peak expiratory flow rate 605 L/min (4.33, 5.21, 82.5% and 643 predicted respectively). Chest x-ray revealed multiple soft tissue density nodules together with reticular shadowing in lower zones (compare the two chest radiographs, Figs 3 & 4).

The following were normal:-

Haemoglobin, white cell count and differential, blood urea, electrolytes, creatinine clearance, mid stream urine, ECG, echocardiogram, serum proteins, liver and

Fig 3. Initial chest X-ray reported as normal.

Fig 4. Chest X-ray 2 months later showing mainly basal distribution of soft tissue nodular and reticular shadowing.

thyroid function tests, calcium studies, radiological studies – barium meal, enema and x-rays of both hands (Fig 5), abdominal ultrasound scan, faecal occult blood negative, and muscle biopsy. Auto-antibodies (double strand anti-DNA, anti-smooth muscle, anti-mitochondrial and anti-parietal cell antibodies) were all negative.

Extractable nuclear antibodies:-

n RNP (nuclear), SM, SS-A/Ro, SS-B/La, Jo-1, rRNP (ribosomal), PCNA, Ku, SL, PL-4, Pm-ScL, PL-7 and PL12 were all negative.

Discussion

A diagnosis of clubbing¹ should be considered with a combination of the following^{1,2}.

1. A bulbous, fusiform enlargement of the distal portion of the fingers and toes.
2. The angle made by the proximal nail fold and the nail plate (Lovibond's angle)³ exceeds 180 degrees.
3. No abnormality of underlying phalanx.
4. There is a noticeable mobility of the matrix as one pushes down and away from the distal interphalangeal joint.

Fig 5. X-ray of hands – no soft tissue calcifications evident.

These criteria were satisfied in this patient, although in systemic sclerosis the degree of nail changes is surprisingly mild even with distinct changes in the pulp of the fingers¹.

The skin rash in systemic sclerosis is typically described as "one or more round or oval, firm, reddish plaques up to several centimetres in diameter that become white or yellow centrally often with lilac coloured telangiectatic border. A guttate, uncommon variant of morphoea has been described, but its status is tenuous since it may be a variety of Lichen Sclerosus Et Atrophicus."⁴

This is characterised by multiple small chalk white lesions primarily involving the anterior chest, neck, shoulders and occasionally other areas of the body⁴.

The skin of the face loses its normal lines, becomes mask-like with telangiectasia, while skin over the fingers (sclerodactyly) shows marked atrophy (shiny tinge) with peri-ungual telangiectasia. Dysphagia occurs in systemic sclerosis in 80% of cases due to abnormal function of the oesophagus⁵. This man did not have CRST Syndrome⁶ (calcinosis cutis, Raynaud's phenomenon, sclerodactyly and telangiectasia) which is a variant of systemic sclerosis. Basal lung fibrosis on chest radiology is typical, but basal nodules have been associated with other autoimmune diseases that form the spectrum, yet histology and immunology were typical of scleroderma⁶. The fact that he has Scl-70 antibodies confirms the diagnosis^{7,8}.

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