SCLEREDEMA - SOME UNUSUAL FEATURES
A Case Report

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Summary

Two cases of adult scleredema has been reported. The unusual electrocardiographic features and association with pulmonary tuberculosis in these cases of scleredema are described. The salient features have been highlighted and relevant literature has been reviewed.

Scleredema is a rare disorder of unknown aetiology which manifests as indurated areas of skin and frequently follows an infectious episode elsewhere in the body. It is known to clear up spontaneously in a matter of months or years.

Till 1975, a total of 225 cases of scleredema have been reported in the literature[1]. We report here two cases with some unusual features.

Case 1.

A 30 year old married muslim female was admitted to the medical wards when she presented with progressive "swelling" of face and upper part of the body of two months duration. She also complained of stiffness of the extremities and shoulder girdle areas for the same duration. History of dyspnoea, palpitation, wound sepsis or any genito-urinary tract disease were denied.

Physical examination revealed a young female, with markedly thickened skin of woody consistency. She appeared to be 'obese' with a facies which lacked expression. Her skin was thickened all over and it showed woody hard consistency with less of marking on the exterior. There was no pitting on pressure. Skin all over was shiny and its colour was normal (Fig. 1). These features were more marked over shoulder girdle, trunk, neck and face. Pinching and folding of skin was impossible. Movement of neck and shoulder regions were restricted. Joints were normal. Systemic examination did not reveal any abnormality. Fundi were normal.

Investigations showed haemoglobin 12.5 gm%, ESR 45 mm for 1st hour. Total and differential leucocyte count, blood urea, serum creatinine, serum electrolytes, total and differential serum protein were all within normal limits. There was no abnormality in glucose tolerance test. Test for L.E. Cell and rheumatoid factors were negative. Roentgenogram of the chest showed enlargement of cardiac silhoutte (Fig. 2). Electrocardiogram taken

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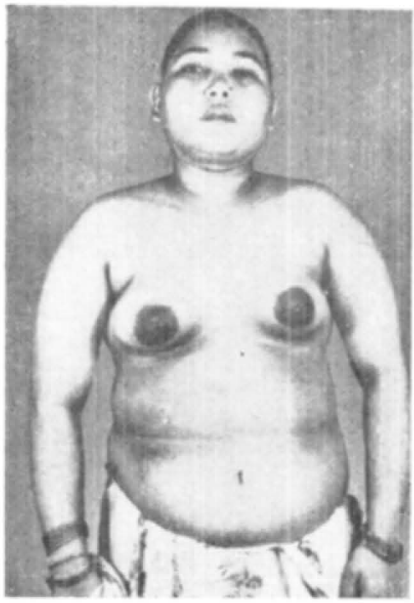

Fig. 1
Case 1: Photograph of the patient showing skin changes.

on admission showed evidence of right ventricular hypertrophy and ST–T–Wave changes in all the leads which were nonspecific (Fig. 3).

Histopathology of skin from the scapular region showed an increase in thickness of dermis, oedema and the latter with splitting and clear spaces between them. Toluidine blue staining showed metachromasia. The changes were typical of scleredema.

Case 2.

A 60 year old hindu male was admitted to the medical wards because of 'hardness' of skin and restriction of movement in the neck and shoulder girdle region of 3 years duration. He also gave history of persistent cough with expectoration of over 20 years duration. History of polyuria, haemoptysis and breathlessness were denied. He had fever occasionally. Along with these the patient complained of pain in joints involving the larger peripheral joints.

Examination revealed tense shiny skin of woody hard consistency which was more marked in the region of neck, shoulder girdles and upper part of arms. Tenderness was absent. Hair growth was normal. Expansion of chest was decreased and there was increased vocal resonance, bronchial breathing and coarse leathery crepitations over the right inter and infrascapular region. Temperature was 99°F and clubbing was absent.

Total and differential WBC count, fasting blood sugar and blood urea

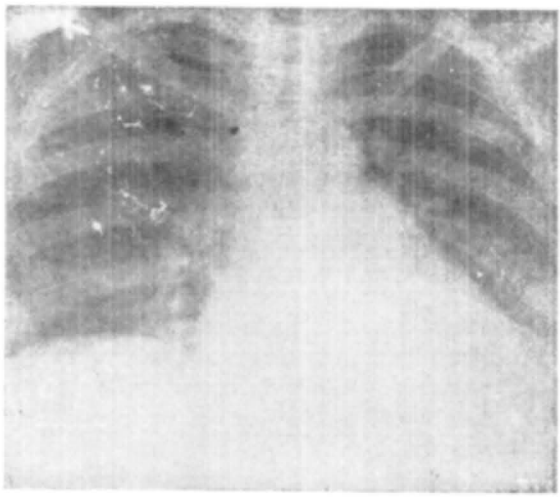

Fig. 2
Case 1: Roentgenogram of chest showing gross cardiomegaly.

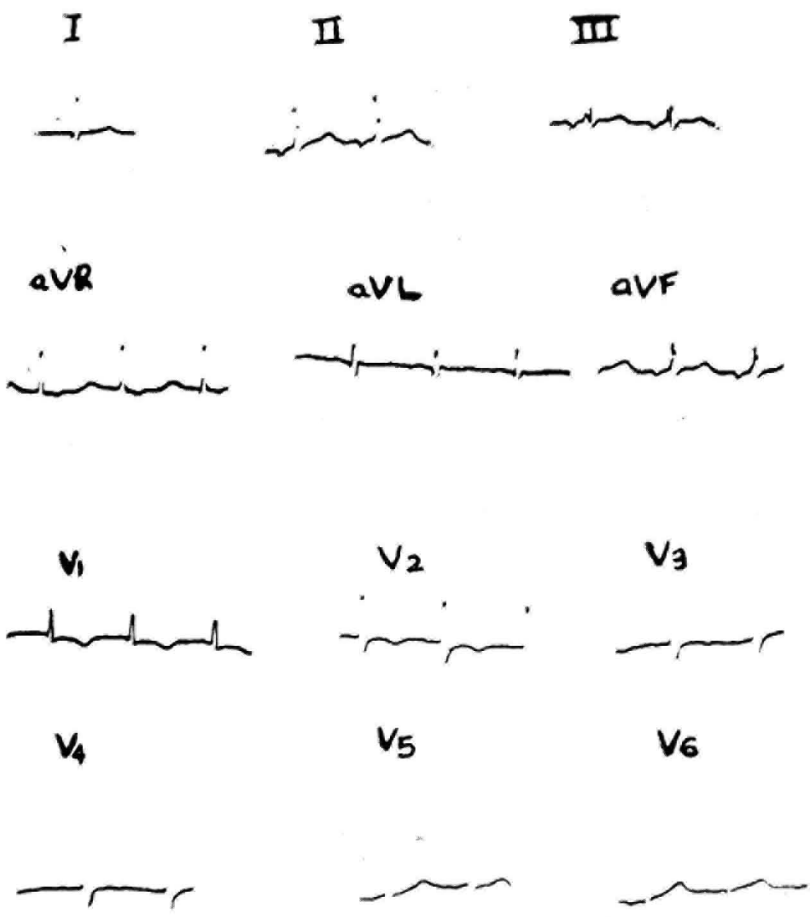

Fig. 3 Case 1: 12 lead electrocardiogram of the patient showing features of right ventricular hypertrophy and T-wave changes.

were normal. X-ray of chest showed bilateral infiltration which was more marked in the right mid zone. The direct smear of sputum was negative for AFB. On culture it yielded a positive growth for AFB. Biopsy of skin was taken from shoulder region and the histopathology was consistent with scleredema (Fig. 4).

Discussion

Though Buschke[2] is credited with the first description of the disease, the entity of scleredema has been known since1752[3]. Classically the patient presents with stiffness of neck and shoulder leading to restriction of movement of these parts. Occasionally skin eruptions have been reported[4]. The disease is known to spare hands and feet[5]. Involvement of external genitalia has been recorded in 6 out of 209 cases reviewed by this author[6]. The cases under discussion had classical features, though in the second case the distal extremities were involved more extensively. Skin eruption or genital involvement were not observed.

Vallee[6] in his review of 107 cases which included four of his own, considered scleredema as a systemic disease by virtue of occurrence of pneumonia, pleural effusion, and hepato-splenomegaly in these patients. Difficulty in protruding the tongue, dysphagia and restricted eye movement are cited as

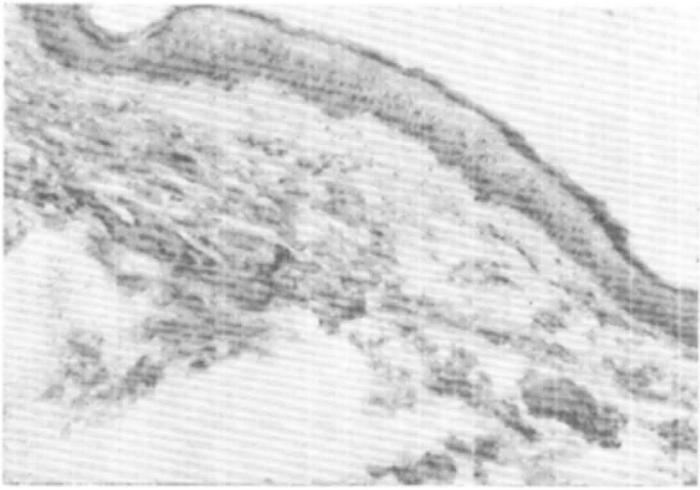

Fig. 4

Case 2: Microphotograph of skin showing, thinning of spiakemis, dermal collagenisation and 'fenestration' (× 10).

evidence suggestive of neuromuscular involvement in this disease[6,7].

Cases with clinical cardiovascular abnormalities are also on record. Pericardial effusion[4,6] diastolic gallop rhythm without heart failure[7] and association of classical rheumatic carditis in this disease[8] are some examples. A variety of electrocardiographic abnormalities have been reported. Reversible RS-T wave changes, prolongation of Q-T interval and d epression of ST-T wave have been reported earlier[7]. Lately Bhargava et al[9] reported right bundle-branch block in a case of scleredema. In the first case described here, apart from radiological enlargement of cardiac silhoutte, there was generalised STi-T changes and evidence of right ventricular hypertrophy in the electrocardiogram. We have not come across any similar report in the literature concerning these changes in relation to scleredema. In view of self limiting nature of this disease exact clinico-pathological correlation of this observation will be difficult to explain. On the same score autopsy and cardiac histopathological studies are not available. Apart from clinical features, histopathology of affected skin shows very characteristic features which confirms the diagnosis. Except for the thinning of epidermis in the second case, the histopathological ehanges were classical.

Occurence of a febrile illness preceding the onset of skin lesion has been observed in 65 to 90% cases[5,10]. Robinow[7] in his review of 76 cases did not find any such infective illness in 23 7% of cases; in the rest a variety of illnesses occured. One of our cases had evidence of pulmonary tuberculosis, an association hitherto not described. Role of this infective illness in the pathogenesis of scleredema is not understood. Many cases of scleredema with diabetes mellitus has been reported[11,12]. This association is often held responsible for nonresolution of the skin lesion contrary to its natural course.

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