

Summary

35 year old female with past history of bronchial asthma, presents with bilateral lower limb oedema up to the level of knee joint, which was persistent throughout the day with no facial puffiness, shortness of breath or abdominal distension. At presentation she was in her fifth month of first pregnancy. There was no history of frothy urine, haematuria or reduced urine output suggestive of renal aetiology. There was no exertional dyspnoea, orthopnoea or paroxysmal nocturnal dyspnoea. There was no alteration in bowel habits. Then she developed bilateral hand swelling and skin thickening over the hands mainly below the wrist joint and gradually the skin thickening progressed with the involvement of forearm, anterior neck and chest and the face. Increased skin pigmentation was noted which was generalized in distribution. She also noticed bilateral, symmetrical small joint pain and swelling with bilateral symmetrical large joint involvement. There was history of difficulty in opening the mouth, but not associated with dysphagia or regurgitation. There was no associated Raynaud phenomenon, hair loss, oral ulcers or photosensitivity. Around the sixth month of pregnancy she developed exertional dyspnoea and orthopnoea. There was no associated fever, cough, haemoptysis, and chest pain or contact history with tuberculosis. There was no history of easy fatigability or history of bleeding manifestations. On examination she had evidence of speckled leucoderma, skin thickening below the elbow joint and over the neck, anterior chest and face. There was bilateral non pitting pedal oedema and increased generalized skin pigmentation. There was no evidence of nail fold telangiectasia, pulp atrophy or pitted scars.

Diastolic blood pressure was elevated. There were no signs of pulmonary hypertension or right heart failure. Auscultation of the lungs did not reveal bibasal fine end inspiratory crepitations in favour of interstitial lung disease associated with systemic sclerosis. There were no features suggestive of active synovitis in small or large joints. Investigations revealed positive ANA and anti Scl70 antibody confirming the clinical diagnosis of diffuse cutaneous scleroderma. Intravenous immunoglobulin was given as the mainstay of treatment along with low dose steroids and hydroxychloroquine. Emergency caesarean section was done due to foetal complications and baby was given neonatal intensive care. Following the delivery she developed renal impairment, mild proteinuria and blood picture evidence of microangiopathic haemolytic anaemia. Since scleroderma renal crisis was an anticipated complication in her scenario with early, aggressive, diffuse cutaneous disease an angiotensin converting enzyme inhibitor was initiated promptly.