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CASE REPORT

Asymptomatic Osteolysis of Ribs and Clavicles in Progressive Systemic Sclerosis

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Abstract: Asymptomatic osteolysis of ribs and clavicles in progressive systemic sclerosis. J. V. Bertouch, T. P. Gordon, D. Henderson and P. M. Brooks, *Aust. N.Z. J. Med.*, 1982, **12**, pp. 627–629.

The association of severe osteolysis of clavicles and ribs in a patient with progressive systemic sclerosis is reported. The disappearance of the clavicles and upper ribs was not associated with any symptoms. The possible causes of this uncommon association are discussed.

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Progressive systemic sclerosis (PSS) is a diffuse connective tissue disorder in which scleroderma-tous skin changes are accompanied by inflammatory, vascular and fibrotic changes in other organs. Typically these changes occur in the gastrointestinal tract, heart, lungs or kidneys. The musculoskeletal system is commonly involved and changes ranging from marginal joint erosions to localised bone resorption have been described.

These changes may occur in the hands, particularly in the terminal phalanges and also in the distal radius and ulna[1, 2], cervical spine[3], mandible[4], clavicle[1, 5] and ribs.[3, 6, 7] In this report a patient with PSS with osteolysis and subluxation of multiple ribs and almost complete resorption of one clavicle is described. These changes, although gross, were asymptomatic.

Case Report

A 42-year-old woman was first seen in 1976 with a history of Raynaud's phenomenon and poor grip strength of both hands with loss of skin creases. She was diagnosed clinically as having scleroderma and investigations including chest X-ray (Fig. 1) were normal at this time. Over the next five years there was rapid progression of the disease. There was marked skin tightening and polyarthralgia and development of digital ulcers in both hands and feet. Areas of subcutaneous calcification developed in the hands leading to ulceration. Treatment with D-penicillamine and colchicine had no effect on disease progression. She was admitted to hospital in February 1981 with several necrotic ulcers over the hands and elbows and symptoms of dyspepsia, easy fatigability and weight loss.

Clinical examination revealed typical changes of scleroderma with the additional findings of finger clubbing, cardiomegaly, and bilateral crepitations extending to the mid-zones of both lung fields. Chest X-ray (Fig. 2) revealed cardiomegaly, a basal reticulonodular pattern, almost complete resorption of the right clavicle, narrowing and resorption of the outer one-third of the left clavicle and dissolution and subluxation of the posterior parts of the 5th to 10th ribs on the right and 4th to 10th ribs on the left. Patchy calcification was seen in the chest wall and also around both shoulder joints. Hand X-rays revealed extensive soft tissue calcification and "pencil-in-cup" deformities. Soft tissue calcification was also present around the right elbow and both ankles and feet.

Pulmonary function studies revealed a forced expiratory volume in one second (FEV1) of 1·2 L (predicted 2·7 L) and vital capacity (VC) of 1·3 L (predicted 3·5 L). FEV1/VC ratio equalled 91%. Maximum mid-expiratory flow rate was 1·5 L/S (predicted 3·2 L/S) and peak expiratory flow was 121 L/M (predicted 376 L/M). Lung volumes and diffusing capacity were not performed as the patient could not hold the mouthpiece in her mouth. A skin biopsy from the right forearm demonstrated a thickened acellular dermis with appendage loss consistent with scleroderma. There was no evidence of vasculitis and immunofluorescence was negative.

Barium studies showed absence of peristalsis in the lower two-thirds of the oesophagus, a hypotonic stomach and dilatation of the proximal duodenum. An ECG demonstrated sinus rhythm and evidence of an old anterior infarction, and echocardiography revealed a dilated, poorly functioning left ventricle consistent with cardiomyopathy. The typical cutaneous changes together with evidence of pulmonary gastrointestinal and cardiac involvement confirmed the diagnosis of PSS. Full blood count showed a microcytic hypochromic anaemia with haemoglobin of 7·3 g/dl and ESR of 45 mm/hour. Biochemistry including electrolytes, urea, creatinine, calcium, phosphate, alkaline phosphatase and liver function tests were within normal limits. Antinuclear antibodies were not detected at any stage of the disease. Immune

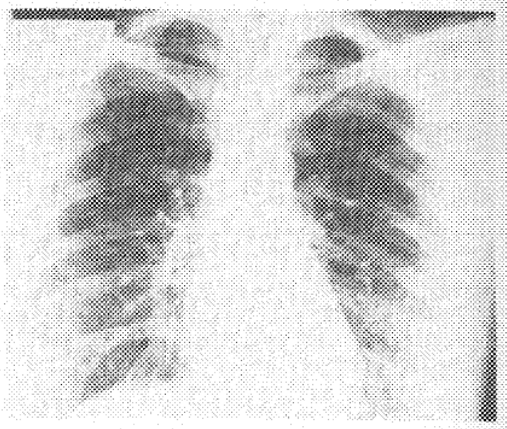

FIGURE 1. Chest radiograph 19.2.76. Normal.

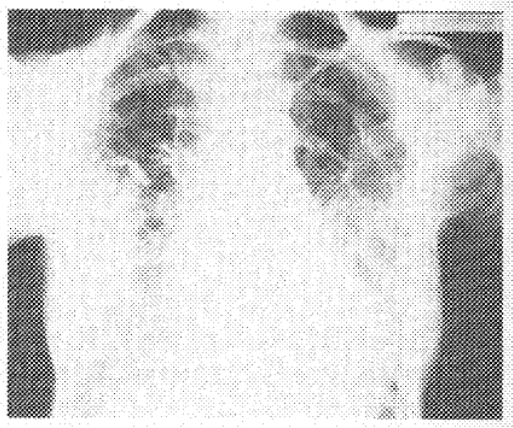

FIGURE 2. Chest radiograph 3.4.81. Dissolution of the right clavicle except for the distal part and tapering of the distal part of the left clavicle can be seen. There is erosion, dissolution and subluxation of multiple ribs on both sides with soft tissue calcification. Increased basal lung markings and cardiomegaly are also present.

complexes measured by a C1q binding technique were slightly elevated at 5·1 U (normal < 2 U).

The cause of the anaemia was presumed to be gastrointestinal blood loss but endoscopic examination could not be performed because of the small oral aperture. Faecal occult blood was not detected. She was discharged on oral iron supplements, cimetidine and diuretics and haemoglobin rose to 11·2 g/dl over the next six weeks.

Two months later readmission was required because of a two-day history of rapidly progressive dyspnoea. Examination revealed atrial fibrillation and pulmonary oedema with congestive cardiac failure. Symptoms were attributed to cardiomyopathy and controlled with bed rest, fluid and salt restriction, digoxin and diuretics.

Two months later a further hospital admission was required with increasing dyspnoea but this episode proved resistant to all treatment and the patient died in June 1981. Permission for a post mortem examination was denied.

Discussion

This case report represents the most severe example of osteolysis in PSS that has been reported. A remarkable feature is that the patient had no symptoms referrable to the chest during the five-year period in which bone resorption occurred. The only symptom noted was the change in contour of the upper chest as the right clavicle disappeared. It is also interesting that serum calcium, phosphate and alkaline phosphatase remained within normal limits throughout the course of the disease despite both osteolysis and widespread soft tissue calcification. Radiologically, the progressive tapering of ribs appeared to be due to resorption of cortical bone. The subsequent osteolysis and subluxation occurred in the posterior portion of each involved rib. Classically the earliest rib lesion seen in PSS is erosion of the posterior superior aspect[8], allowing differentiation from inferior notching due to vascular impression. The changes in the right clavicle are unusual both because of the extent of the osteolysis and because the distal part, although markedly tapered, is still present. It is much more common in PSS for the distal clavicle to be eroded and tapered with sparing of the proximal portion. These features were seen in the left clavicle in this case. Additionally, osteolysis was seen in areas quite removed from joints and there was no radiological evidence of periosteal new bone formation.

The mechanism of bone resorption in PSS is not known. Bone loss in the phalanges is usually ascribed to reduced blood flow secondary to intimal proliferation and spasm of small vessels. Haverbush *et al.*[3] proposed ischaemia as the likely cause of osteolysis of the cervical spine in their case report but found no vascular abnormality on biopsy. However, an ischaemic mechanism for bone resorption may be supported by the report of avascular necrosis of the femoral head in a patient with PSS who had not received corticosteroids.[9] Histological examination of the intercostal or internal thoracic vessels in PSS for evidence of vasculitis has not been performed to our knowledge. Rib erosions are also well recognised in rheumatoid arthritis, and are considered to be due to scapula pressure or to bursitis in the region between the scapula and thorax.[10, 11] The extensive lesions seen in this case make this explanation unlikely in PSS.

The radiological appearance of the right clavicle resembled that seen in another condition variously known as vanishing bone disease, massive osteolysis or Gorham's disease.[12, 13] This is characterised by complete disappearance of part or all of a bone or bones in the absence of any systemic disease. In this condition the characteristic finding in tubular bone is tapering of the margin of the lesion to a pencil point. There is no evidence of any metabolic or endocrine disturbance. Biopsy of affected areas shows replacement of bone by angiomatous tissue and the arteriovenous shunting may cause high output cardiac failure. This is particularly interesting in view of the well-known vascular lesions of skin and other organs in PSS. Histological examination of involved bone and its vascular supply in future cases of PSS may help to elucidate the cause of this uncommon feature.

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