

LETTERS

Multiple rib infarcts: a rare form of osteonecrosis in antiphospholipid syndrome

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Osteonecrosis (ON) is a well known morbidity of several rheumatic diseases, usually affecting the hip, knee, shoulder, and ankle. The involvement of multiple or atypical sites by ON has been reported, especially in association with antiphospholipid antibodies (aPL). Rib infarction has been reported as a painful crisis of sickle cell disease, but has not been reported in rheumatic diseases. I describe here the first case of rib infarcts which developed in a patient with mixed connective tissue disease (MCTD) and secondary antiphospholipid syndrome (APS).

CASE REPORT

A 36 year old woman with hand oedema, Raynaud's phenomenon, and polyarthritis of both hands was diagnosed as having MCTD 3 years ago. She had deep venous thromboses in both legs 2 years ago. Laboratory tests showed an antinuclear antibody titre of 1/2560, IgG anticardiolipin antibody 65.42 GPU (normal <15 GPU), and anti-U1RNP antibody (1/1280). Secondary APS associated with MCTD was diagnosed, and treatment with low dose steroid, aspirin, and warfarin was prescribed.

Recently, she presented with a 15 day history of diffuse, bilateral chest pain. Physical examination disclosed tenderness on multiple ribs. Laboratory findings showed an increased leucocyte count of $11.5 \times 10^9/l$, decreased platelet count of $85 \times 10^9/l$, and an increased erythrocyte sedimentation rate of 36 mm/1st h. Coagulation tests showed an activated partial thromboplastin time of 72.4 seconds, and other tests were within the normal range. Immunological studies showed C reactive protein of 125 mg/l (normal <50) and IgG anticardiolipin antibody 45.63 GPU. Other immunological tests including anti-Sm, anti-dsDNA antibody, cryoglobulins, homocysteine were normal or negative.

Bone scintigraphy showed non-uniform uptake, with areas of decreased and increased uptake in the anterior and lateral aspects of multiple ribs, bilaterally (fig 1). Histological analysis confirmed multiple ON of the ribs (fig 2). She was prescribed with simple analgesics in addition to previous drugs. Her chest pain improved dramatically over the following week and her disease was stable on follow up.

DISCUSSION

In this report, I describe a rare case of multiple rib infarcts in a patient with secondary APS who had been managed with low dose glucocorticoid. Two interesting points about this case should be emphasised. The first is the pathogenesis of ON. ON has obvious association with various systemic conditions, including sickle cell disease, prolonged glucocorticoid treatment, alcohol abuse, and Gaucher's disease, and several factors, such as vascular thrombosis, mechanical defects, cellular damage, embolisation, changes in introsseous pressure, or trauma, have been proposed in its pathogenesis.¹ For rheumatological diseases, a pathogenic relationship between aPL and ON has been proposed on the basis of clinical observations of patients with APS who

developed ON in the absence of glucocorticoid treatment.^{2,3} Several studies showed a correlation between aPL and ON in patients with systemic lupus erythematosus,^{4–6} and Belmonte *et al*⁷ suggested a possible association of aPL with ON in patients with HIV infection.

Multiple rib infarcts in this case strongly suggest that a systemic process may be the underlying pathogenesis. Corticosteroid use and the vasculitic process can be considered to be the causative factors in this case. However, ON in this patient was controlled with a relatively small dose of corticosteroid, and clinical disease was relatively inactive when ON was diagnosed. Whether aPL are directly responsible for this condition or whether they are present as bystanders remains controversial. But, owing to the hypercoagulable potential of aPL, vascular occlusion or thrombotic vasculopathy related to the aPL was also thought to be the underlying pathogenetic mechanism. Thus, the prothrombotic potential of aPL and the above described clinical features of this patient suggest that aPL are directly implicated in the aetiopathogenesis of ON.

The second point which should be emphasised is the atypical presentation of ON affecting the ribs bilaterally. This is an unusual, first described presentation of ON developing on multiple ribs bilaterally. Rib infarction has been reported rarely as a painful crisis of sickle cell disease and associated with vaso-occlusive phenomena.⁸ Although the involvement of multiple or atypical sites (that is, vertebral body and lunette bone) has been reported in association with aPL or catastrophic APS, rib infarcts have not been reported in rheumatic diseases. I suggest that rib infarcts may be an unusual complication of rheumatic diseases with prothrombotic potentials, especially APS.

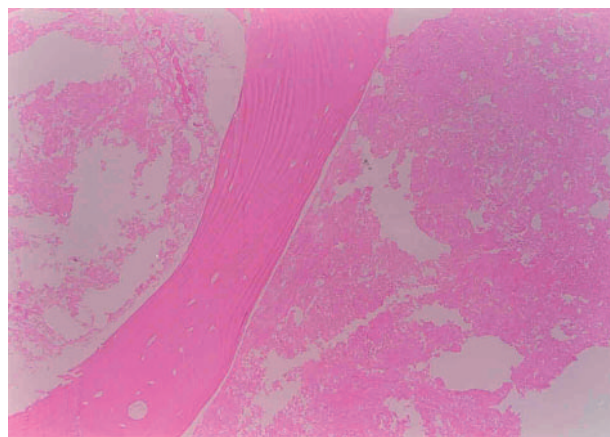


Figure 2 Histological examination of trabecular bone shows the signs of bone death. The trabeculae are preserved, but the lacunae are empty and the marrow is dead (haematoxylin and eosin $\times 100$).

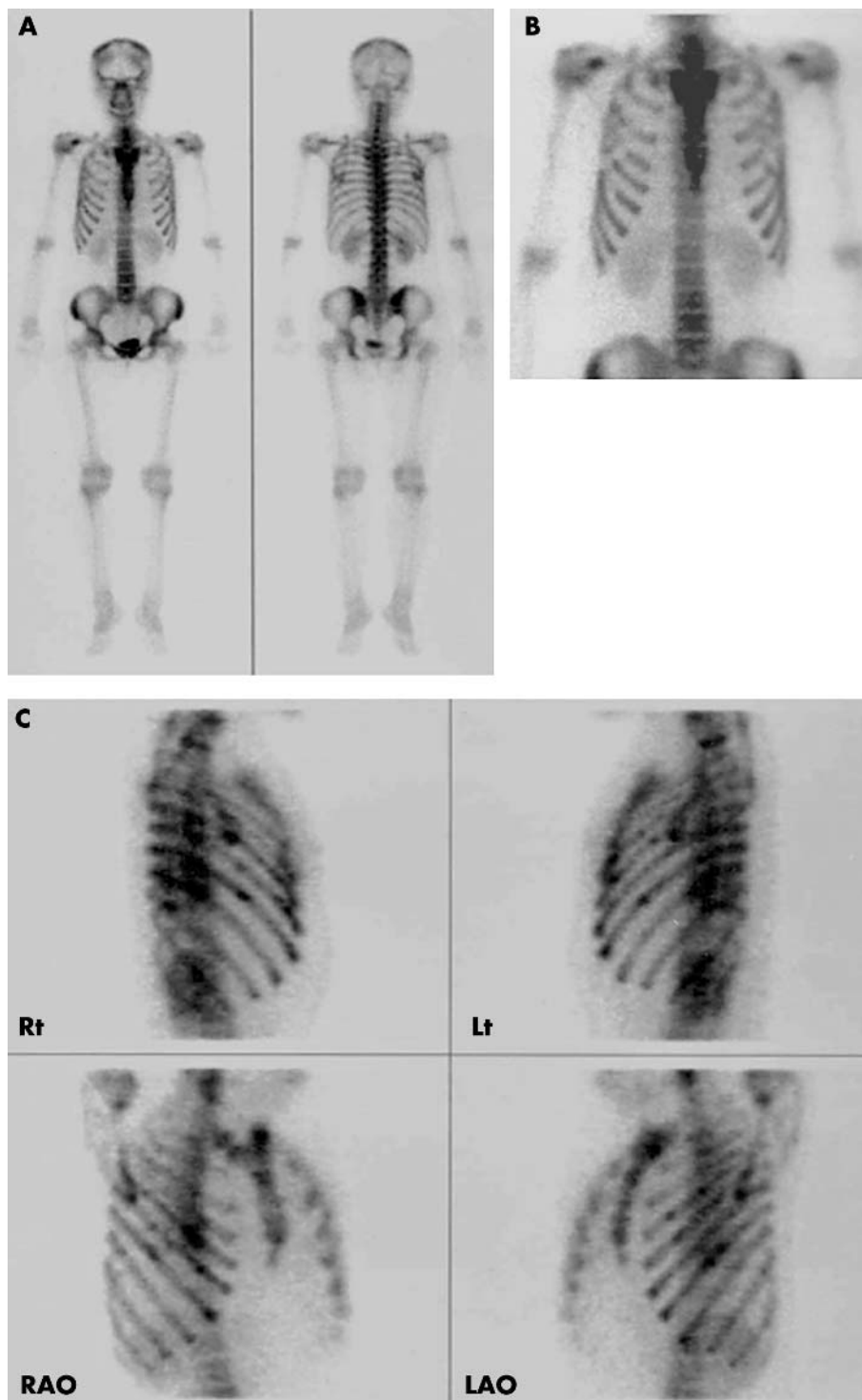


Figure 1 Bone scintigrams of whole body (A), anterior (B), lateral and oblique (C) views of the chest show multiple areas of decreased and increased uptake in the ribs, consistent with bone infarction.

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