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CASE REPORT

Unusual serosal calcifications in CREST syndrome

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Introduction

CREST syndrome is a limited form of systemic sclerosis comprising calcinosis, Raynaud's phenomenon, oesophageal dysmotility, sclerodactyly and telangiectasia. The calcinosis is usually confined to skin and subcutaneous tissues and is found most commonly in the hands. We report an unusual case in which calcinosis involved all three serosal membranes (peritoneum, pleura and pericardium). To our knowledge such a pattern of calcification in CREST syndrome has not been reported before.

Case report

A 77-year-old Asian woman presented with limited cutaneous scleroderma and low back pain. She had Raynaud's phenomenon involving both upper limbs for 4 years, dysphagia, and sclerodactyly along with digital infarction and facial telangiectasia. There was no history of previous asbestos exposure or past tuberculosis. Autoimmune profile revealed positive anti-nuclear antibody (titre 1:320; finely speckled pattern); negative anti-centromere antibody, anti-double-stranded DNA, anti-cardiolipin and lupus anticoagulant antibodies. A radiograph of both hands showed terminal tuft resorption of most digits. Minimal peri-capsular soft-tissue calcifications were seen involving the left second and third metacarpo-phalangeal joints.

Radiographs of the lumbar spine and pelvis revealed scoliosis with degenerative changes, but the most striking abnormality was multiple round calcific densities of variable size overlapping the spine and pelvis (Fig. 1). To determine the exact location of these calcifications, an abdominal computed tomography (CT) examination was performed. It clearly showed the calcifications were related to peritoneum and small bowel mesentery (Fig. 2). Plaque-like subcutaneous calcifications were also noted in both gluteal regions (Fig. 2(a)). High-resolution chest CT showed no evidence of interstitial lung disease, however, a few small round calcific densities were noted along the visceral pleura of the major fissures in both the lungs (Fig. 3(a) and (b)). Faint plaque-like pericardial calcifications (Fig. 3(c) and (d)) were also present. A supine barium swallow showed oesophageal hypomotility associated with mild dilatation and hold up.

Discussion

Progressive systemic sclerosis (scleroderma) is a collagen vascular disease with progressive sclerosis of connective tissue and the vascular system. It has two subsets: diffuse or limited cutaneous forms. Diffuse cutaneous scleroderma is a rapidly progressive disease with widespread skin involvement,

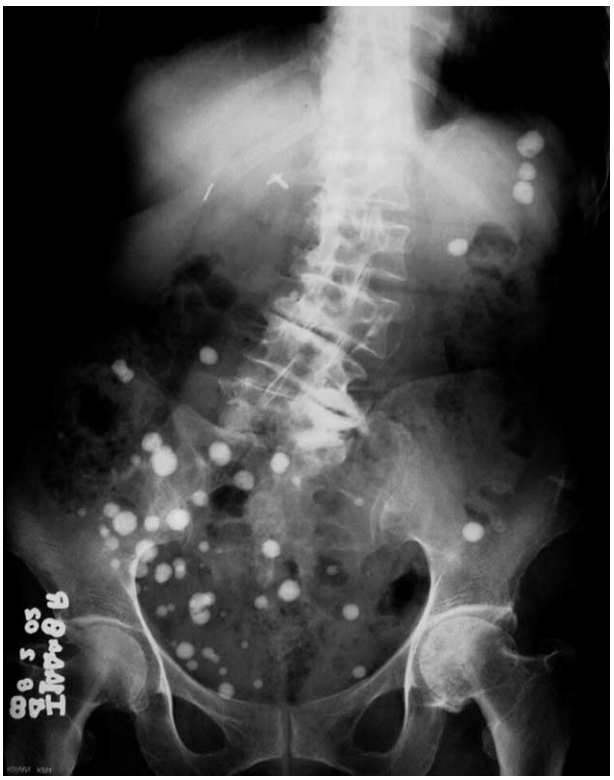

Figure 1 Abdominal radiograph shows multiple nodular calcifications of variable sizes. Incidental findings include degenerative change with scoliosis and cholecystectomy clips.

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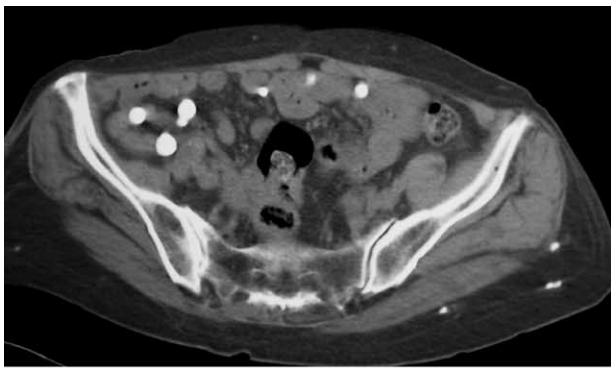
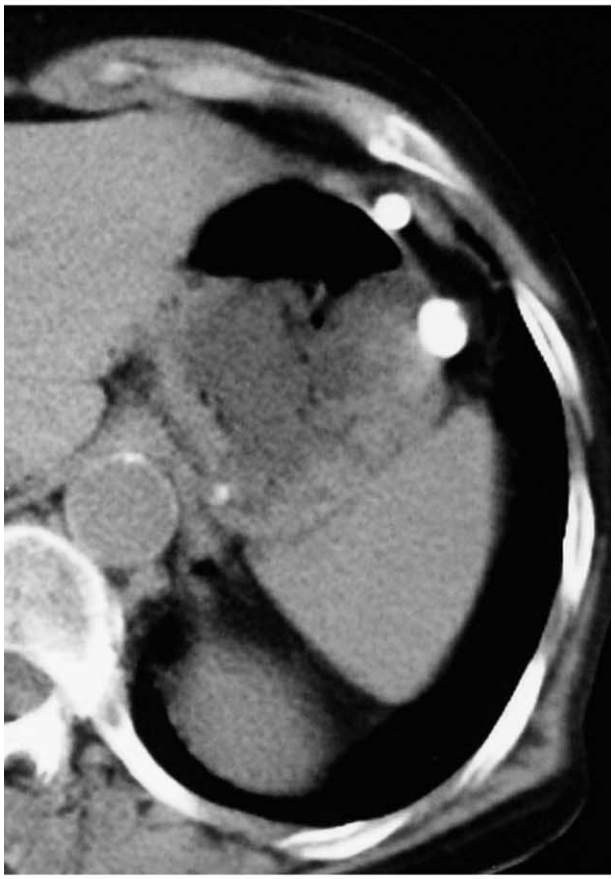
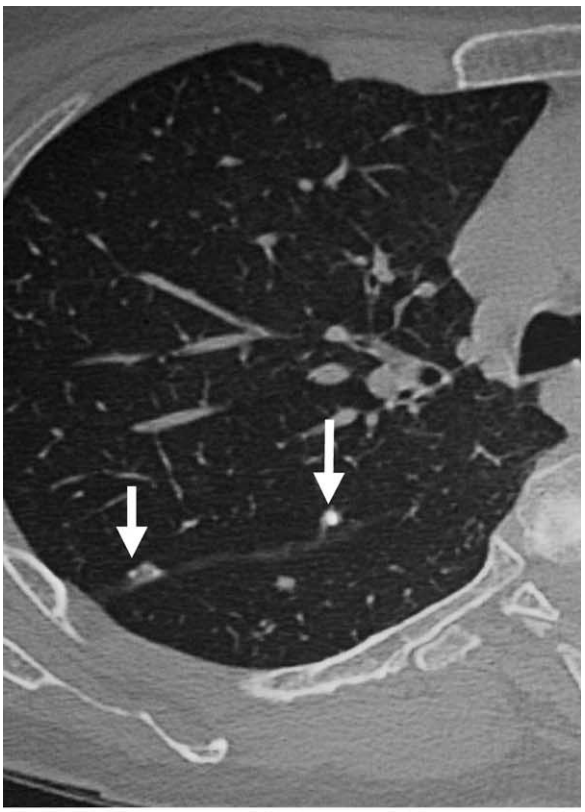


(a)

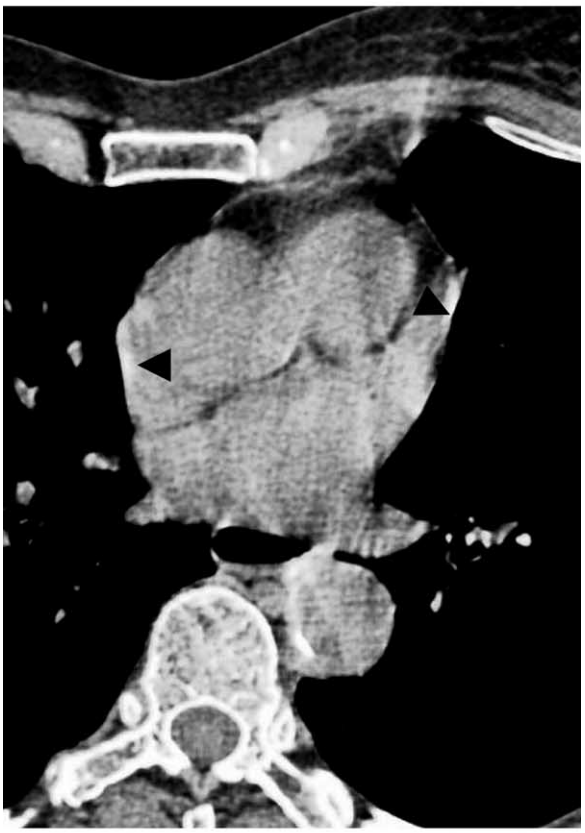


(b)

Figure 2 Axial CT images of pelvis (a) and abdomen (b) show rounded nodular calcifications involving visceral peritoneum and small bowel mesentery and plaque-like subcutaneous calcifications in the left gluteal region.



(a)



(b)

Figure 3 Axial chest CT images show small nodular calcifications along the right major fissure (arrows) and pericardium (arrowheads).

early development of visceral abnormalities and shortened survival. In contrast, limited cutaneous scleroderma is associated with slowly progressive skin changes usually restricted to distal forearms, hands, fingers and face. This limited cutaneous subset frequently has features of CREST syndrome. Calcinosis, an integral part of CREST syndrome, is found most commonly in soft tissues and periarticular tissues. These calcifications are usually in the form of small amorphous deposits in the fingertips and around the joints of hands, feet and in the skin of forearms, but can be more extensive and form large clumps of amorphous, subcutaneous calcific deposits or widespread thin calcific plaques in skin and subcutaneous tissues.

The mechanism of soft-tissue calcification in scleroderma is not well understood. It is generally accepted that it is caused by local tissue factors, particularly microvascular insufficiency and local tissue injury, rather than abnormal calcium and phosphorous metabolism.[1] Although various degrees of subcutaneous calcification are a common feature of scleroderma or CREST syndrome, rare atypical patterns of calcification have also been described, including extensive brain parenchymal calcification involving basal ganglia, dentate nuclei and subcortical white matter[2] and spinal calcifications leading to canal stenosis.[1]

Cardiac involvement in scleroderma may manifest as pericardial disease, myocardial disease or a conduction system abnormality. Whereas pericardial involvement has been found in 33–71% of scleroderma patients in various autopsy series, the clinical symptoms of pericarditis were recorded in only 7–20%.[3] Fibrinous pericarditis, adhesions or effusions may represent pericardial disease in these patients. Pericardial effusions have been reported in 33% of patients with CREST syndrome.[4] One CREST syndrome patient developed sudden worsening of his congestive heart failure ascribed to calcific constrictive pericarditis.[5] The present patient had thin, plaque-like pericardial calcifications but had no symptoms or signs suggestive of pericarditis. Such a pattern of pericardial calcification in the absence of pericarditis has not, as far as we know, been reported earlier in scleroderma patients.

Calcifications seen on the abdominal radiographs are commonly localized to peritoneum and lymph nodes. In tuberculosis, the calcified lymph nodes are usually distributed along the root of mesentery and retroperitoneum and appear as clustered, round to oval, mottled or popcorn shaped. Stippled calcifications can be seen in previously irradiated lymph nodes in Hodgkin's disease in the retroperitoneum. Other causes of nodal calcification are malignant tumours, including neuroblastoma, cervical carcinoma, testicular embryonal cell carcinoma, ovarian germinoma, papillary cystadenoma and adenocarcinoma of gastrointestinal tract.[6] Diffuse peritoneal calcifications may be seen as a sequel to tuberculous peritonitis, *Armillifer armillatus* infection or peritoneal metastasis especially from ovarian carcinoma. In *A. armillatus* infestation calcified nymphs are seen in lungs and peritoneum, usually in the right upper quadrant of the abdomen overlapping the liver region and appear as coiled calcifications measuring 4–7 mm on plain radiography. Other causes of peritoneal calcification are rupture of primary hydatid cyst in the peritoneal cavity, multiple phleboliths in haemangiomas, and sclerosing peritonitis following long-term peritoneal dialysis.

Virtually any process that can cause pleural thickening can go on to calcify,[7] but in practice calcification is mostly due to infection, haemorrhage or asbestos exposure. Nodular tiny calcific bodies seen 'hanging' from the visceral pleura in the present case are unlike any of the known causes that result in pleural calcification.

Although various clinical patterns of calcification have been described in CREST syndrome, the present patient represents an unusual case in which the calcification involved all three serosal membranes of the body.

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