Calcinoid pus presenting as an abscess in a patient with limited cutaneous systemic sclerosis

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DESCRIPTION

Systemic sclerosis (SSc) is a rare autoimmune, connective tissue disorder of unknown aetiology.¹ The prevalence of the disease is 19 cases in a million and it is more common in women aged 30–50 years. SSc is characterised by widespread fibrosis of the skin, various internal organs or both of varying degree of severity. In limited cutaneous systemic sclerosis, previously called CREST Syndrome (calcinosis, Raynaud's phenomenon, oesophageal dysmotility, sclerodactyly, and telangiectasia syndrome), the skin tightening occurs distal to the elbows or knees and the face, while in diffuse SSc, the skin of the proximal extremities and trunk is involved. Current treatment for SSc is directed towards managing complications and providing symptomatic relief. In addition to disease-modifying treatments, surgical intervention may be required in infected lesions.

Calcinosis in SSc results from the deposition of insoluble calcium salts in the skin and subcutaneous tissues. Local trauma, vascular hypoxia and chronic inflammation leading to dysregulated mesenchymal stem cell differentiation and a bone-forming microenvironment have been

Figure 1 Plain radiograph of the left hip and thigh showing extensive calcification centred over greater trochanter region extending into the surrounding soft tissue and no gross abnormalities in the underlying cortical bone.

Figure 2 Contrast-enhanced MRI T1/T2 weighted of the pelvis showing a large fluid collection measuring 4.6×3.6 × 3 cm, extending from the femur to the skin.

proposed to the formation of calcinosis.² These heterotrophic ectopic calcific deposits may lead to ulceration, sinus formation and infection that contribute to SSc-associated morbidity. Clinical and radiological diagnostic imaging with surgical excision of symptomatic calcinoid deposits remain the mainstay of treatment.³

A 69-year-old woman with limited cutaneous SSc was referred to our orthopaedic unit with a 5-month history of progressive dull aching pain and swelling in her left thigh. She was not on steroids or immunosuppression. Her current medication comprised vasodilators, antihypertensives and symptomatic treatment for dry eyes and dry mouth. She was apyrexial. Clinical examination demonstrated a large diffuse swelling in the left upper thigh with overlying erythema. It was tender to touch with pain-free ipsilateral hip joint movements. The plain radiographs revealed extensive calcific deposition centred over the region of the greater trochanter extending into the surrounding soft tissues; however, there was no gross abnormality in the underlying cortical bone (**figure 1**).

The blood markers showed a raised C reactive protein level of 70 mg/L, a white cell count (WCC) of $8.5 \times 10^9/L$ and an erythrocyte sedimentation rate of 78 mm/hours. T1 (precontrast and postcontrast) and T2 MRI revealed a large fluid collection measuring 4.6×3.6 × 3 cm, all the way down, lateral and posterior to the proximal femur with a rim enhancement (**figure 2**). On the suspicion that this was an abscess, the patient was taken to theatre for incision and drainage.

Surgical exploration revealed a large collection of dense white fluid (**figure 3**). Microbiological examination showed high leucocytes but no microorganisms. Extended cultures were negative. The presence of carbonated hydroxyapatite crystals confirmed the diagnosis of liquefied

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Figure 3 Specimen pot with fluid collected on surgical exploration and sent for microbiological examination.

calcific pus. The wound healed with secondary intention, over a period of several weeks, leaving behind a puckered scar. The patient felt significant symptomatic relief.

This case highlights the clinical manifestation of a rare disorder. An inflammatory collection of calcinoid pus can present as an abscess. Familiarity with radiological features and complementary imaging helps determine the extent and localisation of the problem. Surgical treatment provides microbiological confirmation and resolution of symptoms.

Learning points

- ▶ Calcinosis in systemic sclerosis (SSc) can mimic an infective process or a tumour.
- ▶ Calcinosis in SSc is a common cause of morbidity and can be found incidentally on radiographs or seen as obvious clinical presentation.
- ▶ Surgical exploration helps in microbiological diagnosis and definitive management of unusual liquefied calcinoid collection.

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