Massive calcinosis cutis associated with primary Sjögren's syndrome

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DESCRIPTION

A 28-year-old Japanese woman presented with subcutaneous nodules on the extensor surface of her elbows and knees, which had been present for 6 years. Radiographs of the knees showed numerous calcifications, up to 13 mm in diameter, in the subcutaneous tissue on the extensor side of those joints, consistent with calcinosis cutis ([figure 1](#)).

The patient's renal function, serum calcium, phosphorus and parathyroid hormone levels were normal, and there was no evidence suggesting a metabolic disorder. Antinuclear antibody (40×, speckled) and anti-Sjögren's syndrome antigen A (SSA) antibody were positive. Although her subjective symptoms of dry eyes and dry mouth were mild, parotid sialography showed stage two salivary gland disease with collections of contrast material measuring 1–2 mm in diameter throughout the gland ([figure 2](#)). There were no signs of other connective tissue diseases (CTD), hence, based on the revised Japanese criteria for Sjögren's syndrome,¹ the patient was diagnosed with primary Sjögren's syndrome (pSS). There were signs of neither

Figure 2 Parotid sialography showing stage two salivary gland disease.

nephrocalcinosis nor of renal tubular acidosis. Treatment with diltiazem was ineffective, and surgical excision of the lesions was performed with no recurrence after 7 months of follow-up.

Calcinosis cutis has been reported to occur in association with various CTD, most commonly with systemic sclerosis and dermatomyositis. There have been two reports of calcinosis cutis associated with pSS in the English literature: one in the fingertips of a 55-year-old woman² and another in the hands of a 74-year-old man.³ However, to the best of our knowledge, calcifications this massive have not been reported in association with pSS before.

Figure 1 (A) Frontal and (B) lateral radiographs of the knees, showing numerous calcifications in the subcutaneous tissue.

Learning points

- ▶ Primary Sjögren's syndrome may be associated with calcinosis cutis.
- ▶ Surgical excision of the lesions may be effective for this condition.

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Competing interests None declared.

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