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Parotid calcification in systemic lupus erythematosis

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Abstract
Calcinosis universalis is an unusual complication of systemic lupus erythematosis (SLE). We describe the case of a 22-year-old patient diagnosed with SLE during childhood, who developed a previously unreported complication of parotid calcification. Medical treatment of calcinosis is often ineffective, whereas surgical intervention completely relieved the patient's symptoms.

Key words: Parotid Gland; Calcinosis; Lupus Erythematosis, Systemic

Introduction

Calcinosis is an unusual complication of systemic lupus erythematosis (SLE). When calcification is localized to small deposits on the extremities or around joints, it is referred to as calcinosis circumscripta, and when generalized, as calcinosis universalis. Although a variety of medical treatments are available to reduce the progression of calcification the overall results are disappointing.

We describe the previously unreported occurrence of parotid calcification complicating SLE that required surgical excision.

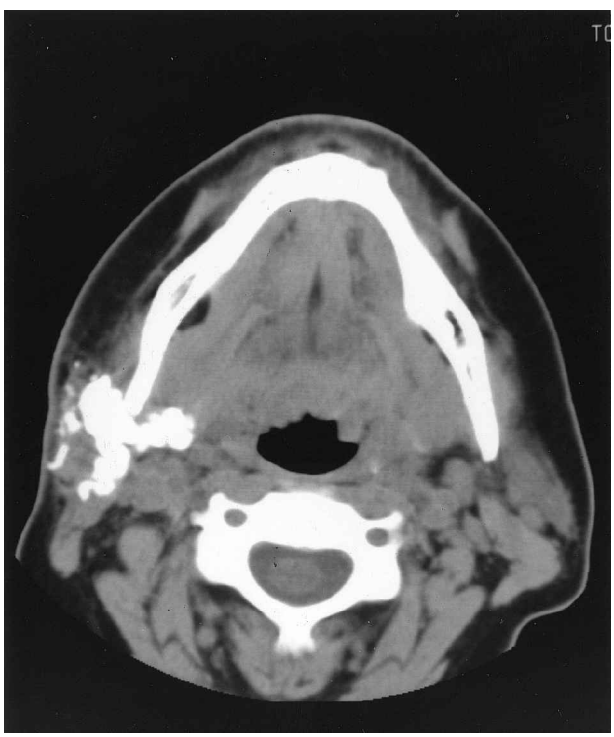

Fig. 1
Computerized tomogram demonstrating calcification with both deep and superficial lobes of the right parotid gland

Case report

A 22-year-old presented to our department with a painful mass in the region of her right parotid gland. She had been diagnosed at the age of 10 years with SLE when she presented with a typical rash across her face, ear lobes and hands, calcific skin nodules, Raynaud's phenomenon and arthralgia. Investigations showed a positive lupus band test, and raised titres of anti-nuclear antibodies (ANA) and p-anti-nuclear cytoplasmic antibodies (pANCA).

A year later she developed intermittent pain and swelling in her right neck associated with a large calcific skin nodule overlying the parotid gland. An open skin biopsy performed in the referring unit demonstrated extensive calcification with tiny remnants of salivary tissue. Unfortunately she developed chronically discharging sinuses along the biopsy scar.

Previous treatment under the care of the lupus team had included hydroxychloroquine, etidronate and diltiazem, but was ineffective.

Examination revealed a 4 cm by 5 cm hard mass within the right parotid gland. Serum calcium (corrected for albumin) and phosphate were normal (2.35, 1.6 mmol/l respectively). CT (computed tomography) scans (Figure 1) demonstrated extensive calcification in both the superficial and deep lobes of the parotid.

In view of the failure of medical treatment, she underwent a subtotal conservative parotidectomy via an extended parotid incision excising the existing sinuses. Gross calcification and infection were noted (Figure 2), and surgery was technically difficult. The facial nerve was identified at the squamo-tympanic fissure and the superficial gland removed. Calcified material was also removed deep to the main trunk, although some parts of the gland were left *in situ* in order to maintain integrity of the nerve. She made an excellent post-operative recovery with a mild marginal mandibular nerve weakness, that has improved significantly with time. Pain was completely resolved for a two-year period. She has recently re-presented with pain in the same region associated with a calcific skin lesion. Injection of Marcain® into the lesion relieved the pain and she will shortly undergo excision of the lesion.

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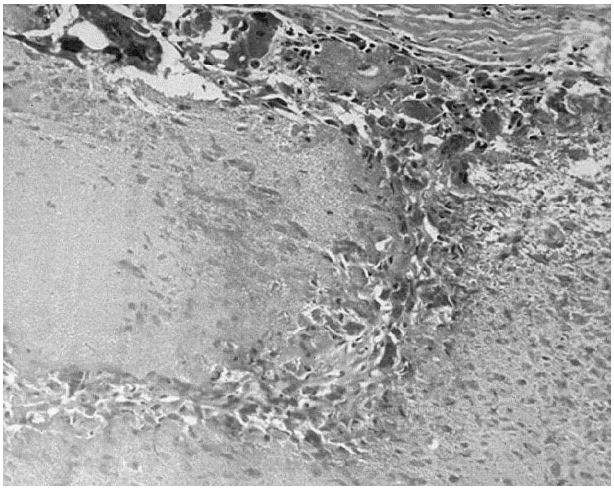

FIG. 2

Photomicrograph showing extensive calcification (in blue) with a peripheral macrophage reaction (H&E; ×200)

TABLE I

CLASSIFICATION OF SOFT TISSUE CALCIFICATION WITH EXAMPLES OF EACH CLASS

Dystrophic calcification	Traumatic
	Inflammatory
Normal calcium and phosphate levels	Degenerative
	Connective tissue disease; SLE, dermatomyositis, scleroderma
Metastatic calcification	Hyperparathyroidism
	Vitamin D intoxication
Abnormal calcium and phosphate levels	Milk alkali syndrome
	Multiple myeloma
Idiopathic calcification	Calcinosis universalis
Normal calcium and phosphate levels	Calcinosis scrotalis
	Tumoral calcinosis

Discussion

Calcification results from the deposition of calcium and phosphate in the organic matrices of soft tissues. It is classified into three main groups; dystrophic calcification, associated with localized or diffuse tissue damage or inflammation; metastatic calcification associated with aberrant calcium and phosphate metabolism; and idiopathic[1] (Table I). Parotid gland calcification is uncommon, except in the setting of sialadenitis. Other reported causes include calcification within tumours (including pleomorphic adenoma, lymphoma and pilomatrixoma), calcium pyrophosphate crystal deposition disease, Sjögrens syndrome and tuberculosis infection.

Clinical features of calcinosis universalis typically include painful plaques or nodules between 0.5–5 cm, symmetrically, distributed over the extremities. The plaques may spontaneously ulcerate leaving a slowly healing sinus discharging chalky material. Many cases of reported 'idiopathic' calcinosis universalis are thought to have subclinical connective tissue disorders, although true idiopathic cases do rarely occur.[2] Calcinosis universalis, although occurring in up to 74 per cent of children with dermatomyositis,[3] is rare in SLE. When associated with SLE, calcinosis more commonly takes the localized circumscripta form and is found in subcutaneous tissues around joints in the setting of long-standing severe disease. Calcinosis universalis was first reported complicating SLE in 1969,[4] and a number of reports have demonstrated calcification of periarticular and muscular sites. Paradoxically, calcinosis predicts a better prognosis in connective tissue disease, but poorer functional outcome.[5]

The physiological mechanism of dystrophic calcification is poorly defined, but occurs in the presence of normal metabolism in damaged or devitalised tissue. Matrix vesicles, membrane-bound structures derived from the endoplasmic reticulum, are thought to act as the initial site of calcification in areas of fibrinoid necrosis.[5] Cell damage is proposed to change cell membrane permeability, allowing calcium flux that results in precipitation of calcium phospate.[6]

Medical treatment of calcinosis is often ineffective, as in the case presented. Etidronate, a bisphosphonate commonly used in the management of Paget's disease of the bone, has been used with some success. However, treatment may result in osteomalacia and pathological fractures, and exacerbation of disease has been noted on treatment withdrawal.[7] A Vitamin K-dependent amino-acid, gamma-carboxyglutamic acid, has been found to act at calcium ion binding sites in bones and soft tissues.[8] Raised urinary levels have been demonstrated in patients with calcinosis and connective tissue disorders.[9] Consequently low dose warfarin, a vitamin K antagonist, has been used in the management of calcinosis. Improvements confirmed on bone scanning are reported in small groups of patients,[10] however this is not accepted in all centres. Corticosteroids have also been used with limited success.

There are few reports of the surgical management of calcinosis universalis in the literature, and these describe involvement of the extremities.[6,11] Excellent symptomatic improvement is described with no reported local recurrence. We have experienced a similar outcome in our case, and agree that surgical excision is of value in this uncommon condition.

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