Case report

Familial calcific peri arthritis

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SUMMARY A family of 4 is described in which both children had calcific peri arthritis affecting the shoulders, and the mother had radiological evidence of peri articular calcification near the left greater trochanter.

Calcific peri arthritis is a condition characterised by acute painful episodes and calcific deposits in the peri articular tissues, most commonly around the shoulder joints.¹ The pathogenesis is poorly understood, and in most cases no obvious local or general cause for these deposits is found. Pinals and Short² described patients with involvement at multiple sites and suggested that it may be a generalised disorder. The frequent soft tissue calcification in patients with renal insufficiency undergoing maintenance haemodialysis would support this hypothesis.³ A familial association in calcific peri arthritis is not as clear-cut as in chondrocalcinosis or gout. So far there have been 3 reports of it,^{4–6} and this is a further report on a family with this disorder.

Case report

A 37-year-old housewife presented with a 4-year history of pain in both shoulders. She had not responded to treatment with nonsteroidal anti-inflammatory drugs. She was otherwise well and had no relevant past medical history. Her mother suffered from 'rheumatic' pains and her brother had recently developed pain in his shoulders. On examination she had severely restricted movement in both shoulder joints. Radiographs showed calcific deposits in the region of the supraspinatus tendon on both sides. Investigations of the sedimentation rate, rheumatoid and antinuclear factors, serum calcium, phosphate, uric acid, and magnesium gave normal results.

Local hydrocortisone injections failed to relieve

her symptoms, and surgery was undertaken to remove the calcific deposits from the left shoulder. At operation the calcific material consisted of a dry, chalky powder adjacent to the supraspinatus tendon. Considerable improvement of symptoms resulted at the operated site.

Her brother, aged 34 and previously well, developed sudden severe pain in his right shoulder. The differential diagnosis at his local hospital where he was admitted was gout and septic arthritis. A radiograph showed calcification in the region of the supraspinatus tendon similar to that of his sister. Investigations were normal except for a high sedimentation rate (40 mm in the first hour). His symptoms failed to resolve with conservative measures, and surgery was carried out. A semisolid substance was easily removed from around the supraspinatus tendon. He too did well post-operatively. Several weeks later, however, he developed pain in his left shoulder. This had a gradual onset and did not prevent him from continuing his occupation. A radiograph demonstrated a similar pattern of calcification.

The parents of the 2 patients were not related. The father never suffered from any rheumatic complaints, while the mother, aged 60, complained of having had pain in her right shoulder for at least 4 years. On examination she had some restriction of movement. X-ray of the involved joint was normal. A radiological survey was carried out on all the family members in a search for further calcification, and the only other abnormality found was calcific deposits near the left greater trochanter of the mother. Biochemical tests, which included plasma glucose, serum urea, sodium, potassium, bicarbonate, chloride, calcium, phosphate, magnesium, albumin, total protein, bilirubin, liver enzymes, and uric acid gave normal results in all the family members.

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Tissue typing revealed the following antigens:

Mother		Father	
A ₂	A ₃	A ₂	A ₂
B ₈	B ₇	B ₇	B ₈
DR ₃	DR ₂	DR ₂	DR ₂

Son		Daughter	
A ₂	A ₂	A ₂	A ₂
B ₈	B ₈	B ₈	B ₇
DR ₃	DR ₂	DR ₃	DR ₂

Discussion

The mechanisms involved in the deposition of different types of crystals in tissues is not yet clear.⁷ In gout a generalised metabolic defect is present. There is, however, no direct evidence of such a defect in either chondrocalcinosis or calcific periarthritis. The former condition has been associated with many different metabolic and endocrine disorders, such as hyperparathyroidism, haemochromatosis, diabetes mellitus, and hyperuricaemia. The significance of this association is not clear, but taken together with the strong genetic element found in some cases of chondrocalcinosis it would support the hypothesis of an underlying generalised abnormality. The only disorders that have been associated with calcific periarthritis are chronic maintenance haemodialysis for renal insufficiency³ and diabetes mellitus.⁸

A familial association in calcific periarthritis is not as clear-cut as in chondrocalcinosis. Cannon and Schmid⁴ described a pair of identical twins with calcific deposits at multiple sites and speculated on the presence of an inherited genetic defect underlying the problem in some patients. Zaphiropoulos and Grahame⁵ described a patient with recurrent calcific periarthritis involving multiple sites. She had no siblings, but 2 of her maternal first cousins had minor calcific deposits in the finger joints, and one of them also had calcification in a dorsal intervertebral disc. Both were asymptomatic. The possibility of a familial element was suggested. The third report is by Marcos *et al.*,⁶ who described the condition in 5 members of the same family, 4 siblings and the mother, and demonstrated the presence of hydroxyapatite crystals in the calcific deposits of one of the 5 patients. Multiple intervertebral disc calcification was found in 4 out of 5 patients, and 3 had, in addition, periarticular calcific deposits. The shoulder joints were involved in 2 of the patients.

In the family we describe 3 out of the 4 members had radiological evidence of periarticular calcifica-

tion. Both children had a similar clinical presentation with bilateral calcific deposits in the rotator cuff giving rise to their symptoms. They had no radiologically apparent calcification elsewhere. The mother had asymptomatic calcific deposits near the left greater trochanter and complained of pain in the right shoulder. The underlying pathology in this joint could not be established, and whether or not there was a pathological process similar to that of her children remains speculative. It could also be argued that the calcification near the left greater trochanter of the mother was a coincidental finding, as this is found in approximately 2% of the normal population.⁹

An association between calcific periarthritis and the HLA system is not yet established. Amor *et al.*¹⁰ reported an increased incidence of HLA A₂ and BW₃₅ in their patients compared with control subjects, but the number of their patients studied was small and the association has yet to be confirmed. Marcos *et al.*⁶ found no association with a particular HLA haplotype among 17 members of the family they described. The results in the 4 members of our family are similarly inconclusive. The father, who was asymptomatic, shared the same haplotypes as the rest of the family.

An inherited genetic factor in some patients with calcific periarthritis seems certain. An association with the HLA system has not yet been established.

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