Systemic Amyloidosis Secondary to Tumoral Calcinosis

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Sir,

Tumoral calcinosis is a rare familial disorder with calcium phosphate precipitation in soft tissues, associated with hyperphosphatemia [1–5].

A 24-year-old Muslim Arab was evaluated for protracted diarrhea, anemia and nephrotic syndrome. At the age of 3, he was diagnosed as suffering from familial tumoral calcinosis, manifested by subcutaneous lumps which occasionally discharged white chalky material, and associated with hyperphosphatemia. Two brothers had the same syndrome. In spite of treatment with aluminium hydroxide and a low phosphorus diet, serum phosphate ranged from 5.3 to 10 mg/dl, mean 6.7 ± 0.3 (SD, $n = 27$). Serum calcium, alkaline phosphatase, creatinine and pH were normal. While the other subcutaneous tumors had disappeared after childhood, one soft-tissue mass which had never opened to the skin and was deeply seated in the left hip gradually increased in size, becoming progressively more prominent (see X-ray, fig. 1). The overlying skin intermittently showed mild signs of inflammation and the patient suffered from recurrent fever with microcytic anemia, thrombocytosis and elevated erythrocyte sedimentation rate. At the age of 22, he developed a severe nephrotic syndrome, with a urinary protein of 18 g/24 h, diarrhea and weight loss. A rectal biopsy was found positive for amyloid in a perivascular pattern characteristic of secondary amyloidosis.

An interesting feature of tumoral calcinosis is that the calcified tumors may evoke local and systemic inflammation such as skin eruptions reminiscent of vasculitis [5], anemia [3], splenomegaly [3], elevated ESR [3, 5] and hypergammaglobulinemia [5]. In our case, chronic inflammation around a large calcified mass in the hip led to the development of secondary amyloidosis, with severe nephrotic syndrome and malabsorption. The pathophysiological mechanism may be similar to the reported association between chronic tophaceous gout or pseudo-gout and amyloidosis [6]. Metastatic calcifications may induce serious systemic signs of inflammation including secondary amyloidosis.

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Announcement

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