ABSTRACT

Three females and one male aged 13 to 25 years and born to non-consanguineous parents presented with tumoral calcinosis. They presented with swellings on the extensor surfaces of their hips, shoulders, and/or elbows, with or without skin ulcers exuding milk-like fluid and scars from previous excisions. Their joint movements were generally free apart from mild limitation caused by skin ulceration or large masses. The masses were made up of calcified material surrounded by a dense fibrovascular reaction and occasional giant cells. Their serum uric acid, blood urea and creatinine, and creatinine clearance levels were all normal, but they had higher serum phosphorus and lower serum calcium levels. Phosphate-binding antacid was prescribed to lower the intestinal absorption of phosphorus, and their diets were adjusted to lower their phosphorus and calcium intake. After 2 years of treatment, their serum phosphorus levels dropped to, or near, normal levels. The lesions resolved completely in 2 of the patients but less so in the other 2 owing to interruption of their treatment. No recurrence, complications, or side effects were noted.

Key words: calcinosis; neoplasms; phosphorus

INTRODUCTION

Tumoral calcinosis is a rare condition characterised by hyperphosphataemia, normocalcaemia and deposits of calcium phosphate, usually adjacent to large joints (such as the extensor aspect of the shoulder, hip and elbow).\textsuperscript{1-5} It manifests in childhood or adolescence as painless, firm, mobile, tumour-like masses in the soft tissue around the joint that may interfere with joint function when large. It is found predominantly in people of sub-Saharan African origin\textsuperscript{3,6,7} Its natural history is not well understood. Inborn errors of the metabolism,\textsuperscript{1} or transport, of phosphorus,\textsuperscript{5,8,9} or a genetically conditioned predisposition of collagen toward calcification\textsuperscript{10} have all been suggested as
possible underlying mechanisms. It is considered a variant of calcinosis universalis, or calcified bursa, or ectopic synovial membrane with surrounding tissue reaction, or degeneration of collagen near the joint, with subsequent calcification. Tumoral calcinosis is usually familial and associated with hyperphosphataemia and recurrence after excision is frequent. Administration of preparations that inhibit absorption of phosphorus diminishes the size of the tumours. We describe successful medical treatment for 4 such cases in one family.

CASE REPORTS

Between January 1997 and January 1999, 4 siblings (3 females and one male) aged 13, 16, 25, and 18 years and all born to non-consanguineous parents presented with tumoral calcinosis (Figs. 1–3). Patients 1 to 3 presented with swelling on the extensor surfaces of their right, or both, hips, with skin ulcers exuding milk-like fluid, with or without scars from previous excision. Patient 3 also complained of low back pain (owing to calcification of the intervertebral disc at

Figure 1  Patient 1: radiographs of the hip (a) before and (b) after treatment and (c) at 8-year follow-up.

Figure 2  Patient 4: radiographs of the elbow (a) before and (b) after treatment and (c) at the 8-year follow-up.

Figure 3  Patient 3: radiographs of the lumbar spine (a) before treatment and (b) at the 8-year follow-up.
L2–3). Patient 4 presented 8 months after the first 3 cases with swelling in both his shoulders, left elbow and hip. All patients had normal renal function, and described the disease manifesting early in their second decade of life.

The nodular masses were of variable size and were hard but movable with respect to bone and skin. Their joint movements were generally free apart from mild limitation caused by skin ulceration or large masses. Radiographically, the masses appeared to be large, calcific, multilocular, cystic structures. Biopsies (done in patients 1 and 2) revealed the masses were made up of calcified material surrounded by a dense fibrovascular reaction and occasional giant cells (Fig. 4). Serum uric acid, blood urea and creatinine, and creatinine clearance levels were all normal, but all the patients had higher serum phosphorus and lower serum calcium levels. The supernatant fluid contained high levels of phosphorus and protein and a moderate level of calcium. The deposit contained a high level of calcium phosphate mixed with a moderate level of calcium oxalate.

The patients were prescribed oral antacid tablets (glycine 180 mg and calcium carbonate 420 mg), 2 tablets 3 times daily, for 3 years. Their diet was adjusted to reduce the phosphorus and calcium intake. Patients 1 to 3 were followed up for 24 months and patient 4 for 16 months; all were followed up again at 8 years. Their serum phosphorus levels fell to, or near, normal levels (Table). Soon after treatment began, their swellings increased in size and became cystic, and the discharge increased for 3 months until the skin ulcers healed. The lesions diminished markedly in patients 1 and 3 but less so in patients 2 and 4 owing to interruption of treatment (military service in patient 4). All patients had a full range of movement in their joints. Patients 1 to 3 stopped the treatment after 2 years and had no recurrence. No complications or side effects were noted.

**DISCUSSION**

Phosphate-binding antacids, together with a diet low in calcium and phosphorus, has been used to lower the intestinal absorption of phosphate and treat calcinosis universalis and tumoral calcinosis. The calcium phosphate complex in tumoral calcinosis is readily exchanged with calcium and phosphate in the serum. Medical management of this condition is preferable because of the metabolic nature of the disease and the frequent recurrence of the lesions after excision.

**REFERENCES**

2. Baldursson H, Evans EB, Dodge WF, Jackson WT. Tumoral calcinosis with hyperphosphatemia. A report of a family with

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**Table**

<table>
<thead>
<tr>
<th>Follow-up (months)</th>
<th>Serum phosphorus level (mg/dl)</th>
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* Normal: 5–6 mg/dl in children, 2.5–5 mg/dl in adults
† Normal: 1.5–6.5 mg/dl in adult females, 2.1–5.6 mg/dl in adult males, 4–7 mg/dl in children