Case Report

Tumoral calcinosis and calciphylaxis presenting in a dialysis patient

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Introduction

Tumoral calcinosis and calciphylaxis are uncommon complications of haemodialysis therapy and are associated with poor phosphate and calcium control. The use of calcium containing phosphate-binding agents in conjunction with high calcium dialysates may exacerbate these conditions by inducing a positive calcium balance. Despite their common aetiology, the two disorders are seldom reported to occur in the same patient. We describe the case of a patient who presented with both tumoral calcinosis and calciphylaxis and subsequently developed devastating vascular complications. This case highlights the morbidity and mortality associated with these diseases, the importance of strict calcium/phosphate control and the dangers of positive calcium balance in dialysis patients.

Case

A 49-year-old male haemodialysis patient presented with painful, swollen shoulders and lesions on his fingertips in July 1997. His primary renal disease was autosomal dominant polycystic kidney disease and he started peritoneal dialysis in October 1990. In July 1991 he received a cadaveric renal transplant, which failed immediately post-operative due to venous thrombosis and he was started on haemodialysis. Despite a superior vena-cava stenosis and two failed radial fistulas, he achieved a urea reduction ratio of >65% with 12 h of haemodialysis each week via a right internal jugular Permacath. However, the patient’s phosphate was persistently elevated above 2.5 mmol/l (normal range 0.8–1.2 mmol/l) despite taking 6 g/day of calcium carbonate as a phosphate binding agent. Serum calcium concentrations were at the upper limit of normal and PTH was 66 pg/ml (normal range 10–55 pg/ml).

Physical examination revealed bilateral shoulder masses (Figure 1) with markedly reduced range of movement. There were ulcerating lesions on the dorsum of his hand and calcific lesions on the distal aspects of his fingers and thumb (Figures 2 and 3). X-rays of his shoulder and hands revealed extensive

Fig. 1. Bilateral shoulder masses due to tumoral calcinosis.

Fig. 2. Areas of superficial ulceration on the dorsal aspect of the hand from calciphylaxis.
soft tissue calcification (Figures 4 and 5). Specific treatment was instigated with increasing doses of non-calcium containing phosphate binding agents (magnesium carbonate 1 g three times a day), withdrawal of vitamin D analogues and increased duration of dialysis against a low calcium dialysate (dialysate calcium 1.0 mmol/l). The new therapeutic regimen stopped the progression of his calcific lesions and his symptoms were controlled with analgesia. Pre-dialysis phosphate concentrations were slightly improved (between 1.8 and 2.2 mmol/l) and serum calcium was reduced to 2.15–2.30 mmol/l (normal range 2.10–2.60 mmol/l).

One year later he received a second cadaveric renal transplant, which was anastomosed to his left external iliac artery. The remaining iliac vessels were noted to be heavily calcified at the time of operation. Two weeks post-transplant he developed a rapidly enlarging left external iliac artery aneurysm, which necessitated a transplant nephrectomy and an iliofemoral bypass. The graft thrombosed 10 weeks later and after the patient declined thrombolysis, a left above knee and subsequent hind quarter amputation was performed. He returned to haemodialysis but developed painful ulcerating lesions on his right leg. The leg ulcers subsequently became infected and he eventually required a right lower limb amputation in July 1999. Histology of a muscular artery, removed at the time of amputation, revealed medial calcium deposition consistent with calciphylaxis (Figure 6). The patient died 4 months later following withdrawal of dialysis.

**Discussion**

Hyperphosphataemia is a common complication of chronic renal failure and is associated with increased mortality [1] and morbidity. Although an elevated calcium/phosphate product is a risk factor for the development of both calciphylaxis and tumoral calcinosis, they are seldom reported in the same patient.

Tumoral calcinosis is characterized by massive periarticular soft tissue deposition of calcium and phosphate and is usually associated with chronic renal failure [2]. Patients usually present with localized swelling, pain and reduced mobility [2,3] most commonly affecting the shoulder, elbow, hand.
and ankle [2,3]. Treatment strategies include tight control of calcium and phosphate levels, parathyroidectomy for hyperparathyroidism, renal transplantation and local excision of calcific lesions.

Calciphylaxis is reported to occur in 1–4% of dialysis patients [4,5] and is characterized by microvascular medial calcification and intimal hypertrophy associated with cutaneous ischaemia and ulceration [5,6]. The initial skin lesions are painful, indurated and violaceous. These lesions usually ulcerate, become necrotic and secondary infection is the main cause of mortality [6]. The distal areas of the lower limbs are most commonly affected and more proximal disease is associated with a worse prognosis [6,7]. Proposed treatments include local debridement of ulcers, control of calcium and phosphate product and hyperbaric oxygen therapy. Some authors have reported a survival benefit from parathyroidectomy [7], although hyperparathyroidism is not a prerequisite for calciphylaxis [8]. The overall mortality associated with calciphylaxis is 30–87% [6].

This case highlights both the vascular and soft tissue complications of poor calcium/phosphate control in haemodialysis patients and the need for non-toxic, non-calcium-based phosphate binders in order to avoid a positive calcium balance in hyperphosphataemic patients. The dose of dialysis delivered should take into account the phosphate balance of a patient in addition to urea kinetics, and dialysis times often need to be increased in order to avoid or treat hyperphosphataemia. Furthermore, the currently prescribed dialysate calcium concentrations ensure a positive calcium balance at a time when the patient is rendered alkalotic by high bicarbonate dialysis. The use of high dialysate calcium concentrations and calcium containing phosphate binding agents may need to be reviewed when considering the calcium balance of individual patients, in order to reduce the risk of extra skeletal calcification.

Conflict of interest statement. None declared.

References


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