Calcinphylaxis, Early Identification and Management: A Report of 2 Cases

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Introduction
Calciniphylaxis, or calcific uremic arteriolopathy (CUA), is a life-threatening calcification of arterioles leading to necrotic infarcts of the skin and subcutaneous tissue (panniculus adiposus) with a high potential to progress to bacterial sepsis and death.

The prevalence of CUA has been reported to be approximately 4.1% in patients with end-stage renal disease (ESRD) with the reported incidence increasing over the past 10 years1. CUA is associated with chronic kidney disease (CKD) and dialysis vintage of 2 years. He was on peritoneal dialysis with end-stage renal disease (ESRD) and a dialysis vintage of 1.5 years. Six months prior to diagnosis he had severe hyperparathyroidism, female sex, Caucasian ethnicity, obesity, diabetes mellitus, time on dialysis, systemic corticosteroid use, hypoalbuminaemia, and corrected calcium phosphorus product >4.4 mmol/L2,3,4.

Risk factors include ESRD, hyperparathyroidism, female sex, Caucasian ethnicity, obesity, diabetes mellitus, time on dialysis, systemic corticosteroid use, hypoalbuminaemia, and corrected calcium phosphorus product >4.4 mmol/L2,3,4.

Here, we present two cases.

Patient Profiles
Patient 1 is a 57 year old Caucasian male with end-stage renal disease (ESRD) secondary to autosomal dominant polycystic kidney disease (ADPKD), and a dialysis vintage of 2 years. He was on peritoneal dialysis and taking calcitriol and calcium acetate. He was on warfarin for hypercoagulability. His serum albumin level averaged 39 g/L in the six months before diagnosis, was 39 g/L at time of diagnosis, and averaged 44 g/L in the nine months following diagnosis. At time of diagnosis, adjusted calcium phosphorus product was 4.9 mmol/L2. His CUA was diagnosed clinically.

Patient 2 is a 47 year old Caucasian female with ESRD secondary to ADPKD, and a dialysis vintage of 1.5 years. Six months prior to diagnosis of CUA, she had severe hyperparathyroidism and underwent parathyroidectomy. Prior to diagnosis she was on calcitriol and calcium acetate. She was on warfarin for venous thrombosis. Her serum albumin level averaged 39 g/L in the six months before diagnosis, was 16 g/L at time of diagnosis, and averaged 23 g/L in the six months following diagnosis. At time of diagnosis, adjusted calcium phosphorus product was 4.9 mmol/L2. Her CUA was diagnosed histologically.

Discussion
Patient 1 underwent the following treatments: He was taken off calcitriol and calcium acetate. Cinacalcet was started for control of hyperparathyroidism. Warfarin was changed to low-molecular weight heparin. The patient was counselled extensively to reduce phosphorus intake and improve nutrition, and to encourage adherence to his medical regimen. In addition to intensive wound care, he was converted from peritoneal dialysis to haemodialysis and continued on sodium thiosulphate for nearly 11 months until his wounds healed.

Patient 2 underwent the following treatments: She had recently experienced parathyroidectomy. Calcitriol and calcium acetate were changed to cinacalcet and sevelamer. Warfarin was stopped. The patient was given prednisone for one month, and two doses of pamidronate. She was counselled to reduce phosphorus intake, and later was started on nocturnal tube feeds, and supplemented with cyanocobalamin (B12), alpha-tocopherol (E), and zinc. Intensive wound care was performed with consultation from plastic surgery. Sodium thiosulphate was incorporated into the patient’s dialysis treatment. The wounds were healing, however, during month six she was re-admitted to hospital with sepsis and died.

In conclusion, the progression of CUA wounds highlights the need for rapid response when early signs appear. Skin biopsy can confirm clinical suspicion but is not without risk of infection and ulceration5,6.

At this time, there are no prospective randomised controlled studies relating to CUA of which we are aware. Research is limited to observational and retrospective studies3. Future research is needed to improve diagnosis and treatment of this devastating condition.

Patient Profiles

Patient 1

Patient 2

Patient 1

Patient 2

Treatment:

Discussion

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Literature Cited