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PHOTOLETTER TO THE EDITOR

Calciphylaxis: a diagnostic and therapeutic challenge

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Abstract

Calciphylaxis is an uncommon necrotizing dermal condition, most often related to end-stage renal disease, associated with secondary hyperparathyroidism. The incidence is 1-4% per year in dialyzed patients. The condition is characterized by microcalcification of small and medium-sized blood vessels, causing cutaneous and soft tissue necrosis. The etiology of calciphylaxis is poorly understood, although many reports highlight the abnormal calcium-phosphate products as an important etiological factor. Calciphylaxis is associated with significant morbidity and mortality with estimated 5-year survival rates of less than 50%. Sepsis is the main cause of death. We report two patients of calciphylaxis treated with surgical debridement and excellent outcome in both cases. (*J Dermatol Case Rep*. 2013; 7(2): 69-70)

Key words:

calciphylaxis, calcium, kidney failure, surgical debridement, treatment, ulcer

Calciphylaxis is a rare vasculopathy that is characterized by ischemia and necrosis due to calcification, intima fibroplasia and thrombosis of dermal and pannicular arterioles. It has been called a "metastatic-like calcification". Usually calciphylaxis affects patients with end-stage renal disease, with an incidence of 1-4% of dialysis-dependent individuals.[1] There are scarce reports of calciphylaxis in patients with normal renal function and with calcium and phosphorus serum levels within normal range.[2] The pathogenesis of calciphylaxis is poorly understood but hyperparathyroidism (primary or secondary to renal failure), diabetes, obesity, warfarin treatment, protein C or S deficiency are well recognized risk factors. Clinically, calciphylaxis presents as purpuric plaques with spontaneous ulceration and necrosis that often progress to deep tissue necrosis. It is associated with significant morbidity and mortality. It is estimated that 5-year survival is less than 50%.[3] Sepsis, originating in the skin lesions is the main cause of mortality.

Figure 1

Extensive lesion of necrotic tissue on the left lower leg surrounded by livedo racemosa-like purpura.

We report the cases of two patients with chronic leg ulcers that where referred to our Department. The first patient was a 63-year-old obese woman with predialysis renal failure and secondary hyperparathyroidism. She was under warfarin therapy due to atrial fibrillation. She presented with an ulcer in the posterior aspect of her left leg, with irregular borders and a necrotic wound bed (Fig. 1).

The second patient was a 61-year-old woman with chronic renal insufficiency on dialysis for several years that presented with a left leg ulcer once again with a necrotic wound bed and rough borders.

Both skin biopsies showed medial calcification of dermal and subcutaneous arterioles (Fig. 2).

Extensive surgical debridement and appropriate wound care resulted in an excellent outcome in both patients. Lesions resolved within 2 months, and no relapse has been seen in 12 months of follow-up.

Calciphylaxis was first reported in 1962 by Hans Selye, who based his reports on the results of serial animal experiments.[4] Still today, it is a poorly understood, difficult to treat, aggressive condition. Outcomes for all recommended therapies remain unreliable, and there have been no prospective treatment trials. In dialysis patients treatment recommendations have included reduction of serum calcium and phosphorus by use of low-calcium dialysis baths, use of pamidronate and parathyroidectomy. Recently attention has turned to newer treatment modalities that can be initiated simultaneously with calcium-PTH axis control, like sodium thiosulfate. The proposed mechanism is that it improves the solubility of calcium in deposits and thereby makes it available for hemodialysis clearance. Prevention of systemic infection is vital. Diligent wound care and avoidance of trauma are imperative features. Debridement and skin grafting are frequent choices in the treatment of these patients, but the role of debridement is controversial.[5] In our patients surgical debridement and appropriate wound care resulted in an excellent outcome.

Calciphylaxis should always be considered in the differential diagnosis of skin ulcerations in patients with any degree of chronic kidney disease. Early diagnosis and treatment of the disease can change its dismal prognosis.

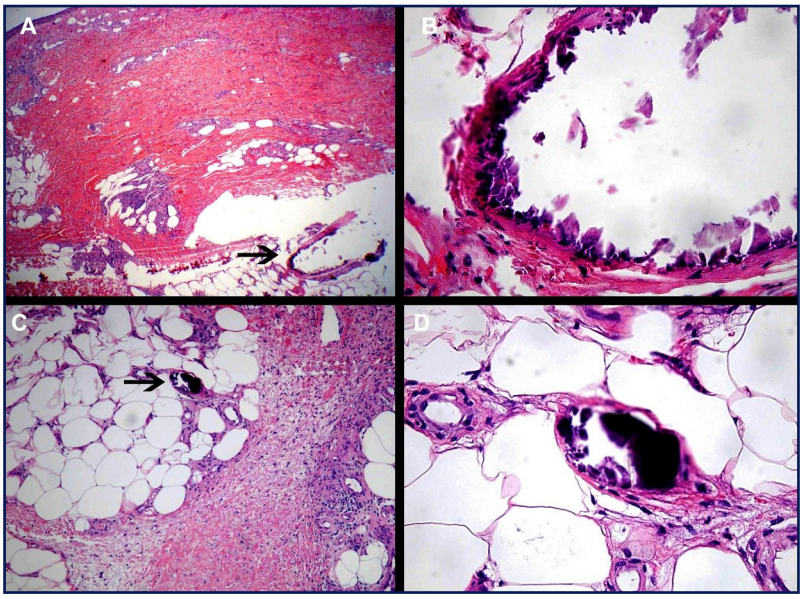

Figure 2

A photomicrograph showing (A) (H&E x40) – arrow shows a subcutaneous arteriole (B) (H&E x400), dense calcium deposits in the vessel wall without evidence of inflammation (C) (H&E x100) – arrow shows an affected arteriole (D) (H&E x400), diffuse and extensive calcification of the media can be seen.

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