

## INTRODUCTION

Calciphylaxis, or calcific uremic arteriolopathy, is a rare medical condition predominantly seen in patients with end-stage renal disease on dialysis. It is characterized by calcification of small and medium-sized blood vessels, leading to skin necrosis and ulcers. Its annual incidence is 35 cases per 10,000 in the United States. Calciphylaxis can be difficult to identify since it is uncommon and has a complex pathogenesis. Hence, before the mortality rate can be decreased, improved understanding and awareness of this illness are required. Prompt and extensive medical intervention is needed to enhance patients' prognosis and quality of life.

## OBJECTIVE

Describe a rare case of a patient with ESRD on peritoneal dialysis who presented with severe progressive calciphylaxis wounds on both lower extremities.

## CASE DESCRIPTION

An 85-year-old male with a past medical history of end-stage renal disease on peritoneal dialysis, prostate cancer status post prostatectomy, esophageal cancer status post resection and radiation, permanent atrial fibrillation on Eliquis, hypertension, hyperlipidemia, and idiopathic peripheral neuropathy, who presented with severe bilateral lower extremity pain. The pain had been present for years but worsened a week prior to presentation.

Upon examination, the patient had diffuse tenderness over the entirety of bilateral lower extremities, some ecchymosis over bilateral distal legs, bilateral motor strength at 2/5 with diminished sensation, and bilateral lower extremity weakness.

Lab results were significant for WBC 16.2, hypocalcemia 7.4, elevated BUN 94, creatinine 9.24, eGFR 5.1, hyperphosphatemia 10.5, PTH 832.3.

US venous doppler bilateral LE was negative for DVT. CTA Aorta/Ileal/Femoral showed stable 4.8 cm ascending thoracic aneurysm, diffuse atherosclerotic but no critical stenosis.

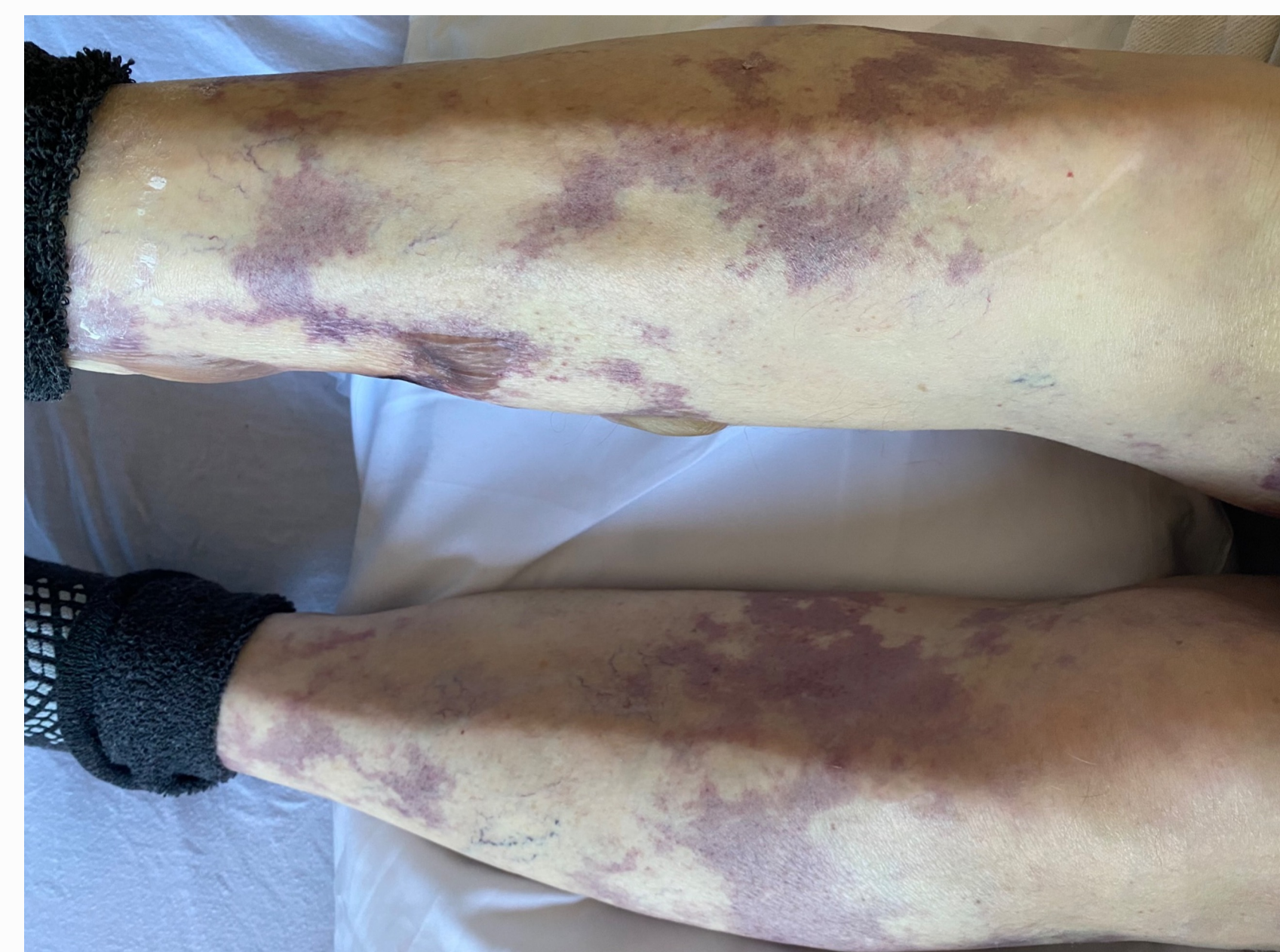
Of note, patient had recent prior admission for left eye vision loss due to suspected giant cell arteritis and a temporal arterial biopsy was done, which showed severe arteriosclerosis with calcifications, and patient was discharged on steroids.

Patient was observed for suspected glucocorticoid induced myopathy due to recent high dose of subsequent prolonged steroids. Statins were also discontinued to rule out statin-induced myopathy.

## CLINICAL FINDINGS

On hospital day 2, patient's bilateral lower extremities worsened, with new ulcerations and eschar formation suspected calciphylaxis. The patient was switched to hemodialysis due to worsening renal function. Pain control, wound care, sevelamer, calcitriol, phoslo, and sensipar were initiated for treatment.

Patient was discharged home on day 5 with close outpatient follow-up.



**Figure 1. Bilateral shins with violaceous patches with surrounding retiform purpura**



**Figure 2. Lateral right thigh with necrotic ulcers, violaceous patches with surrounding retiform purpura**



**Figure 3. Medial right leg with central eschar, formation of blisters, and surrounding retiform purpura**

## DISCUSSION

Exact pathogenesis of calciphylaxis is not fully understood, but it is believed to involve a combination of factors, including hyperphosphatemia, hypercalcemia, and vitamin D dysregulation. Patients with end-stage renal disease are at particular risk, as they often have chronic kidney disease-mineral and bone disorder (CKD-MBD), which can result in these metabolic abnormalities.

In CKD-MBD, the kidneys cannot effectively excrete phosphate, which leads to hyperphosphatemia. This, in turn, can cause a decrease in serum calcium levels, leading to secondary hyperparathyroidism and bone resorption. The resulting increase in serum calcium levels can lead to soft tissue calcification, including the calcification of blood vessels.

A combination of clinical findings, laboratory testing, and histologic examination is essential for diagnosis. Skin biopsy remains as the gold standard for diagnosis, with characteristic findings including arteriolar calcification and thrombosis, ischemic necrosis, and inflammation. Skin lesions associated with calciphylaxis are quite variable in appearance. Firm calcified subcutaneous lesions, which are tender to touch in patients on dialysis, should raise the possibility of calciphylaxis. Pathological and radiographic findings alone are not diagnostic of calciphylaxis and should be clinically correlated. Other tests, such as serum calcium, phosphate, and parathyroid hormone levels, can help establish the diagnosis and identify underlying metabolic abnormalities.

Treatment options include pain control, wound care, calcium-lowering agents, and hemodialysis, as some literature suggests an increased risk of calciphylaxis with peritoneal. Medical therapies for calciphylaxis are limited, and there is currently no consensus on the optimal pharmacologic approach. Sodium thiosulfate, a calcium chelator that improves outcomes in some patients, is often used as a first-line treatment. Other agents, such as bisphosphonates, cinacalcet, and systemic corticosteroids, have also been used with varying success. Surgical debridement and hyperbaric oxygen therapy may also be considered in certain cases.

## CONCLUSION

Clinical awareness and a multi-disciplinary approach in patients with ESRD at increased risk of developing calciphylaxis, especially when presenting with lower extremity pain, skin necrosis, and ulcers. Further research is needed to better understand calciphylaxis's pathogenesis and identify effective treatment strategies.

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