

Calciophylaxis: An Uncommon Culprit for a Common Presentation

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INTRODUCTION

Calcific uremic arteriopathy, or calciophylaxis in patients with ESRD, is the calcification of arterioles and capillaries in the dermis and subcutaneous adipose tissue leading to ulcerative skin lesions.

It is a rare disorder with an estimated 50% 6-month mortality and 1-year survival rates between 45-55% which presents with skin ischemia and necrosis.

Infection is the leading cause of mortality in calciophylaxis and poorer prognosis is associated with advanced disease at the time of diagnosis.

CASE BACKGROUND

HPI: 86-year-old female with 6 weeks of failed outpatient treatment for an enlarging RLE wound with severe pain and increasing purulent, foul-smelling drainage. Two areas of necrotic, non-ulcerated induration on her left medial thigh and calf appeared one week before hospitalization

PMH: class I obesity, CKD Stage V, COPD, atrial fibrillation, hypertension, hypothyroidism, hyperlipidemia, and T2DM

Relevant Meds: amlodipine, furosemide, levothyroxine, losartan, metolazone, rosuvastatin, and warfarin

Exam: 4x5 cm area of tender induration on the right medial thigh with an open erythematous and necrotic eschar with violaceous non-ulcerating skin changes on left thigh (Fig 1)



Fig.1 Bilateral LE wounds on admission

Factors in red should trigger a high index of suspicion for calciophylaxis

LABS AND IMAGING

Imaging: Arterial flow was intact in lower extremities bilaterally with no signs of advanced vascular disease

Labs: Anemia, leukocytosis, elevated creatinine, supratherapeutic INR, hyperparathyroidism, hypercalcemia, and hyperphosphatemia

Histopathology: Superimposed infection and increased risk of bleeding precluded skin biopsy

Ruled out: Atherosclerosis, cholesterol embolization, warfarin necrosis (chronic warfarin treatment), endarteritis obliterans, and vasculitis

MANAGEMENT

- ✓ Broad-spectrum antibiotics, sevelamer, STS, cinacalcet, NOAC, and sodium thiosulfate initiated
- ✓ Discontinued Vitamin D, warfarin, and calcium
- ✓ Risk factor reduction with dietary counseling, peritoneal dialysis optimization
- ✓ Wound care with improvement in wounds

DISCUSSION

Calciophylaxis should be strongly suspected in patients with ESRD or advanced CKD presenting with painful nodules, nonhealing ulcers, or necrosis with eschars in areas of high adiposity.

Other risk factors include obesity, history of warfarin use, hyperparathyroidism, hyperphosphatemia, and hypercalcemia.

The diagnosis of calciophylaxis relies almost solely on a careful history and physical exam with a limited role of skin biopsy.

Management is heterogeneous and centers on risk reduction with discontinuation of warfarin, iron, calcium, and vitamin D; weight management, sodium thiosulfate treatment, calcium, and phosphate-lowering treatment, and appropriate wound care.

Calciophylaxis is easy to misdiagnose, especially in the absence of characteristic cutaneous findings. Therefore, it is important to consider calciophylaxis as one of the top diagnoses for necrotic skin wounds.

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