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IMAGE | LIVER

Calciphylaxis in a Patient With Alcoholic Cirrhosis

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Case Report

A 38-year-old woman with alcoholic cirrhosis was admitted with a recurrent lower extremity rash. She had presented 1 year prior with a history of alcohol abuse, abdominal pain, fatigue, and a violaceous rash over the abdomen and proximal lower extremities. A skin biopsy showed multiple small vessels with calcification of the outer wall suggesting calciphylaxis. The patient was treated with sodium thiosulfate 25 mg intravenously 3 times weekly and serial wound debridement. She had symptom improvement within 3 weeks and resolution of the rash. One year later, the patient presented with recurrence of the painful violaceous rash (Figure 1). She denied any alcohol use over the last year. She had not been receiving albumin infusions or blood transfusions, and her body mass index (BMI) was 22 kg/m^2. Evidence of a healed skin graft was noted on her left thigh, with surrounding tender punctate erythema. Her calcium, phosphorus, parathyroid hormone, creatinine, and protein C and S levels were normal. An abdominal CT was significant for findings consistent with cirrhosis. The patient was started on sodium thiosulfate therapy and local wound care, and responded well without recurrence to date.

Calciphylaxis is a rare, life-threatening syndrome characterized by cutaneous ischemic necrosis secondary to vascular calcification. It is seen almost exclusively in end-stage renal disease, but has been associated with cirrhosis in 9 case reports. The pathogenesis of calciphylaxis is unclear, but commonly described risk factors include female sex, obesity, protein C and S deficiency, corticosteroid use, albumin or blood transfusions, and high calcium-phosphorus product.[1] Clinically, calciphylaxis presents with tender violaceous skin mottling that progresses to blackened regions of eschar formation and non-healing ulceration. This usually occurs on the fat-containing tissues of the trunk, genitals, and medial thigh.[2] Mortality from calciphylaxis has been estimated at 52%, mostly secondary to superimposed sepsis.[1] Management strategies include lowering the calcium-phosphorus product, anti-inflammatory therapy, and aggressive wound care. Sodium thiosulfate, an emerging therapy for calciphylaxis, acts by binding calcium and decreasing reactive oxygen species.[2] Our patient is notable for not exhibiting many of the previously identified risk factors for calciphylaxis. Further studies are necessary to assess factors that pose an increased risk for calciphylaxis in patients with liver disease. Given the high mortality of this condition, prompt recognition and treatment in cirrhotic patients presenting with rash is essential.

Figure 1. Painful violaceous rash on lower extremities due to calciphylaxis.

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References

1. Nigwekar SU, Wolf M, Sterns RH, Hix JK. Calciphylaxis from non-uremic causes: A systematic review. *Clin J Am Soc Nephrol.* 2008;3(4):1139–43.
2. Sowers KM, Hayden MR. Calcific uremic arteriolopathy: Pathophysiology, reactive oxygen species and therapeutic approaches. *Oxid Med Cell Longev*. 2010;3(2):109–21.