Painless Scrotal Ulcers Become Something Unexpected: A Rare Case of Scrotal Calciphylaxis

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Background

Calciphylaxis is a rare vascular disorder that presents with painful skin necrosis due to calcium accumulation in skin and adipose tissue.

Risk Factors 1

• Chronic Kidney Disease
• Hyperparathyroidism
• Long-term Hemodialysis

Epidemiology

• 6 month survival of 50% 1
• ↑ Prevalence in US in HD patients 2

Typical Presentation 3,4

• Painful cutaneous red indurations and eschars
• Nodules which progress to ulcers
• Skin necrosis

This disease carries a high mortality burden which further increases the need to recognize different atypical presentations of this disease.

Case Presentation

A 68-year-old male with a past medical history of ESRD on HD, diabetes, peripheral vascular disease, and neuropathy, presented to the hospital with altered mental status (AMS) and left foot necrosis. After being evaluated by vascular surgery and podiatry, below the knee amputation of the left leg was performed with an unremarkable recovery in the hospital.

Physical Exam:

• Painless mid scrotal 5 cm wound
• No crepitus or fluctuance; Nontender to palpation
• Ca: 8.7mg/dL (normal: 8.5 to 10.2 mg/dL)
• PO4-: 7.3mg/dL (normal: 2.5 to 4.5 mg/dL) ↑
• PTH of 148 pg/mL (normal: 14 to 65 pg/mL) ↑

Results:

US: calcifications in the scrotal tissue
CT pelvis: extensive calcification of the arterial system.
Scrotal biopsy: severe skin and soft tissue necrosis with acute and chronic inflammation; no calcium deposits were seen.

Diagnosis:

A clinical diagnosis of calciphylaxis was made given that the eschar raised concerns for calciphylaxis along with the patient’s comorbidities of secondary hyperparathyroidism, ESRD, and his nonadherence to HD.

Patient Outcome:

Treatment with sodium thiosulfate improved outcome with an eventual discharge. Five days later, the patient was readmitted due to AMS and died due to a cardiac arrest.

Discussion

This case highlights the atypical presentation of painless ulcers as seen in this patient with calciphylaxis. Although the skin necrosis is typical of this disease, the lack of pain perception despite such a severe condition in our patient is noteworthy.

Furthermore, the current literature provides data on several cases of penile calciphylaxis; however, scrotal calciphylaxis is seen less frequently and has little to no data on diagnosis and treatment when the presentation is painless.

This case is an atypical presentation of painless eschars. Therefore, a high degree of clinical suspicion in patients with ESRD on HD and T2DM is needed to effectively reach a diagnosis of calciphylaxis. The high rate of mortality and morbidity coincide with the risk factors associated with this condition. Early recognition of calciphylaxis in a patient with no pain despite the prevalence of ulcerations warrants a closer look into their comorbidities to identify this disease on the differential as it is critical to start treatment early to prevent devastating outcomes.

Conclusion

This case is an atypical presentation of painless eschars. Therefore, a high degree of clinical suspicion in patients with ESRD on HD and T2DM is needed to effectively reach a diagnosis of calciphylaxis. The high rate of mortality and morbidity coincide with the risk factors associated with this condition. Early recognition of calciphylaxis in a patient with no pain despite the prevalence of ulcerations warrants a closer look into their comorbidities to identify this disease on the differential as it is critical to start treatment early to prevent devastating outcomes.

References