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

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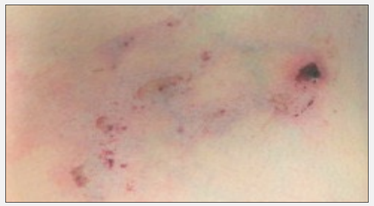
Background

Calciphylaxis is a rare vascular disorder that presents with painful skin necrosis due to calcium accumulation in skin and adipose tissue. Most commonly, cutaneous manifestations of calciphylaxis are seen on the buttocks, thighs, extremities.

Risk factors for this disease include chronic kidney disease, hyperparathyroidism, long-term hemodialysis and diabetes. [1] Several studies show an estimated six-month survival of fifty percent and an increasing prevalence in US in hemodialysis patients. [2] Typically, the disease presents itself with painful cutaneous red indurations, nodules which progress to ulcers, skin necrosis and eschars that are tender to palpation. [3]



Very few cases have presented unconventional locations of these calcified eschars and ulcers such as the penis and scrotum. The current treatment approach for cutaneous manifestations range from wound care, pain management and other supportive care.

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. A few days after the initial presentation, a painless mid scrotal wound and eschar with sloughing was noted on physical exam.

Physical Exam:

- **Nontender, violaceous mid-scrotal** 5 cm eschar
- No crepitus or fluctuance noted

Results:

Lab	Patient Value	Normal Range
Calcium	8.7 mg/dL	8.5 to 10.2 mg/dL
Phosphate	7.3 mg/dL↑	2.5 to 4.5 mg/dL
Parathyroid hormone	148 pg/mL ↑	14 to 65 pg/mL

- 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 12.5g titrated up to 25g 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 presents the atypical presentation of painless ulcers as seen in this patient. 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 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.

Conclusion

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 presence 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

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