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CASE REPORT

Penile Calciphylaxis: Misdiagnosis of a Fatal Condition

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Abstract

Penile calciphylaxis is a rare complication most often occurring in patients with end-stage-renal-disease on dialysis. The condition is associated with a high mortality rate. Misdiagnosis of this life-threatening condition is common. We describe a case of a 41-year-old male with end-stage-renal disease on hemodialysis who suffered multiple admissions for a penile wound. He was misdiagnosed repeatedly leading to significant morbidity. The patient elected hospice care. This case report is intended to bring awareness to physicians to consider calciphylaxis in patients with end-stage-renal-disease on dialysis presenting with painful skin lesions.

Keywords

Calciphylaxis, Kidney disease, Dialysis, Misdiagnosis

Introduction

Calciphylaxis, most often seen in end-stage-renal-disease patients on dialysis, usually presents as refractory and excruciating pain, erythematous skin lesions, and non-healing ulcers that can progress to necrotic plaques. Risk factors include female sex, Caucasian ethnicity, time on dialysis, certain medications, hypoalbuminemia, autoimmune diseases, hypercoagulable states, obesity, malignancy, diabetes, hepatic disease, hyperphosphatemia, and hypercalcemia [1]. Calciphylaxis is a rare but life-threatening and commonly mis-diagnosed condition. Early diagnosis is crucial to providing treatment and preventing morbidity and mortality. This case report highlights the differential diagnoses that can lead to misdiagnosing potentially fatal calciphylaxis.

Case Report

41-year-old male with medical history of end-stage-renal-disease secondary to type I diabetes on hemodialysis three times a week. He presented with fluid overload due to missed dialysis sessions. During that hospitalization, the patient complained about painful penile lesion. Gonococcal and Chlamydia testing was negative and there was no evidence of gangrene or infection. He was referred to outpatient urology follow up. Urology concluded that it was likely a frenulum tear from extreme edematous state and patient was given topical lidocaine and antibiotic cream. Patient's pain was manageable for two weeks before he returned to the hospital with unbearable pain, erythema, and swelling of the penis. Pertinent lab results included WBC 14.5 × 1000, Hgb 8.4 gm%, normal calcium, phosphorus, and lactate levels. Blood cultures were negative and penile wound cultures grew mixed flora. CT without contrast showed a diffusely edematous penis without findings of focal abscess. Urology evaluated the patient, and he was started on antibiotics and local wound care for possible penile cellulitis. Patient had no improvement for two days. Patient failed to respond to antibiotics and developed a necrotic lesion on the glans of the penis. A final diagnosis of calciphylaxis was made. A multi-disciplinary approach involving nephrology, urology, internal medicine, and palliative care was instituted. The patient was educated on surgical options and the high mortality rate of penile calciphylaxis. The patient decided to go home with hospice due to unbearable pain and recurrent hospitalizations in the last year.

Discussion

Penile calciphylaxis has a 6% incidence rate [2] with mortality rates as high as 70% in 6 months [3]. The pathophysiology of calciphylaxis is poorly understood.

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Elevated calcium, phosphorus, and parathyroid hormone levels have been linked to the development of calciphylaxis as mentioned by Westphal, et al. [1]. However, calciphylaxis can occur despite normal levels of calcium, phosphorus, and parathyroid hormone, which occurred with our patient. Misdiagnosis of calciphylaxis is quite common and occurs about 73% as stated by Rrapi R, et al. [4]. Cellulitis is the most common misdiagnosis accounting for 31% as detailed by Gabel, et al. [5]. Other mimickers include warfarin skin necrosis, vasculitis, cholesterol emboli, or peripheral vascular disease [4,6]. Meegada, et al. and Killeen, et al. [7,8] detail cases in which treatment for calciphylaxis was delayed due to initial misdiagnoses of cellulitis. Misdiagnosis can lead to exposing a patient to unnecessary medications which can worsen calciphylaxis and delay initiating essential treatment. This is detailed in the case report by Al Yacoub, et al. [9], in which calciphylaxis was initially misdiagnosed as erythema multiforme/toxic epidermal necrolysis. Patients with end-stage-renal-disease on dialysis are most affected by calciphylaxis due to the erratic bone-mineral patterns. Early recognition of this condition is crucial to providing care through medications, pain management, wound debridement, or surgery. As reported in the case series by Sijapati, et al. [10], early diagnosis and intervening lead to better outcomes. Therefore, it pertinent to include calciphylaxis as a differential in end-stage-renal-disease patients on dialysis who present with localized cellulitis-like areas or open skin ulcers accompanied by excruciating pain.

Declaration of Interest

None.

Conflict of Interest

Each author states that he or she has no associations that pose a conflict of interest with the article.

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Consent

Informed consent was unable to obtained due to patient's demise. Multiple attempts were made to contact next of kin without success.

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