Penile Calciphylaxis: The Use of Radiological Investigations in the Management of a Rare and Challenging Condition

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A B S T R A C T

Penile calciphylaxis is a rare phenomenon of penile necrosis observed in patients with hemodialysis-dependent end-stage renal failure. Multiple treatments have been proposed including conservative management, surgical debridement and penectomy; yet, the prognosis remains extremely poor. Here, we describe a patient with protracted resolution of dry gangrene of the glans, which failed conservative management of wound care and pain management. Radiological studies revealed extensive calcification of abdominal aorta and branching vessels including the penile arteries. Due to intolerable pain, the patient required total penectomy. Earlier surgical intervention guided by findings on radiological studies may improve quality of life in this population.

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Introduction

Penile calciphylaxis, also known as calcific uremic arteriolopathy (CUA) of the penile arteries, is a rare phenomenon occurring mainly in patients with end-stage renal disease and diabetes.1 It is characterized by medial calcification with intimal hypertrophy and small vessel thrombosis.2,3 A recent literature review by O’Neil et al examined all case reports of penile calciphylaxis and found a total of 18 patients (mean age 54.1; range 35–72). They calculated a 69% mortality rate with a mean follow-up of only 5.5 months; thus, this is likely an underestimate of the true mortality rate.4 Although several options have been proposed, there is no standardized management for penile calciphylaxis.

Penile calciphylaxis has been managed either conservatively or surgically, per severity of symptoms and extent of ischemic changes. Conservative management of penile calciphylaxis consists of normalizing serum calcium and phosphate levels, pain management, and administration of sodium thiosulfate (STS), hyperbaric oxygen, appropriate antibiotics and wound care.2,5 Penile calciphylaxis can be treated surgically with wound debridement but often necessitates partial or total penectomy to adequately remove gangrenous tissues until healthy tissue is encountered. However, penectomy has not been shown to have a survival benefit and can be psychologically traumatic.1 Given the high morbidity and mortality of this devastating condition, elucidating effective management strategies is crucial.

Here we present the role of radiological studies in managing a case of penile calciphylaxis, with a protracted course despite conservative management and resolution after total penectomy.

Case presentation

A 46-year-old African American male presented to clinic with a firm, tender, violaceous glans penis. His other medical issues included hypertension, diabetes and hemodialysis-dependent end-stage renal failure. The patient had been on dialysis for 5 years, including prior peritoneal dialysis. Serum calcium and phosphate were 10.7 mg/dL and 6.0 mg/dL. Intact parathyroid hormone (PTH) level was 1096 (normal 15–65 pg/mL). Serum creatinine was 1.4. His medications included doxazosin, furosemide, insulin, lisinopril, losartan, metoprolol and nifedipine. On examination, he had a circumscribed phallic with dry gangrene at the distal half of his glans with no erythema, fluctuance or drainage (Fig. 1a and b). The shaft and base of the penis were minimally tender.
The patient was counseled about conservative and surgical management options. He opted for conservative management with wound care and pain management. Urine was diverted via a suprapubic tube. He only produced minimal urine at baseline. Over subsequent months, the glans continued to atrophy and separate from the healthier penile tissue. The ischemic necrotic glans was expected to slough off at which time the remainder of the penile tissue would heal. In the meantime, he was referred to the pain clinic. Five months after presentation, the patient requested penile amputation for intolerable pain. Penile Doppler ultrasound showed no arterial flow (Fig. 2a) in the penis with extensive calcifications throughout the penile arteries and minimal flow in the dorsal vein. Computed tomography (CT) scan showed air in the penile soft tissues reflecting gangrene up to 6.8 cm from the glans (Fig. 2b), as well as extensive vascular calcification (Fig. 2c). He underwent an uncomplicated total penectomy with penoscrotal urethrostomy. A dry surgical field was noted, with very minimal blood loss, due to the severely obliterated vasculature (Fig. 1c). Histopathology confirmed calcific uremic arteriolopathy (Fig. 1d and e).

**Discussion**

We report a case of severe penile calciphylaxis, in which dry gangrene failed to resolve with conservative management. Due to extensive penile vessel calcification, absent penile blood flow, and ischemia evident on radiological studies, total penectomy was performed.

Given the poor prognosis of calciphylaxis, prompt recognition and treatment is necessary. Several therapeutic options exist, including: conservative management with wound care, antibiotics, analgesics, and medications to reduce serum and tissue calcium content; and surgical management with penectomy.

Radiological studies including penile Doppler ultrasound, CT, and MRI offer assistance in penile calciphylaxis management as per suggested flow chart (Fig. 3). Penile Doppler ultrasound may...
Figure 2. a) Penile Doppler ultrasound showing longitudinal view of right base of the penis, demonstrating lack of flow in cavernosal artery (arrow); b) Pre-operative non-contrast CT scan (sagittal view) showing gas in penile tissue (arrow); c) Pre-operative non-contrast CT-scan (axial view) demonstrating extensive calcification of the cavernosal arteries (arrow).

Figure 3. Suggested flow chart for incorporating radiological investigations in the management of penile calciphylaxis. Note that computed tomography (CT) is well suited to visualize vascular and soft tissue calcifications and gas within necrotic tissues, while magnetic resonance imaging (MRI) typically does not visualize calcifications or gas, but may demonstrate the extent of soft tissue damage, infection and abscess.
be performed initially as a non-invasive tool to assess vascular patency and blood flow in the penile vessels. CT could be performed subsequently as the most sensitive modality to assess the extent of vascular and soft tissue calcification, necrotizing soft tissue infection and/or ischemia with presence of air. It is noted that while ischemia is best illustrated with contrast, this would ideally be avoided in this patient population. MRI may be useful as a tertiary adjunctive modality to delineate the corporal anatomy and extent of ischemia and necrosis without the use of contrast.

Obtaining imaging early on may be beneficial in determining if a patient is a candidate for conservative therapy or a prompt surgical intervention would be more appropriate. Given that our patient did not improve after 5 months of conservative management, he most likely had extensive or progressive vascular calcification, with significantly compromised flow to the penis. An early ultrasound and CT scan may have helped to determine the extent and severity of dry gangrene. It may have elucidated that total penectomy was inevitable and surgery could have been performed earlier. This may have potentially prevented months of pain and poor quality of life.

**Conclusion**

For presentations of penile calciphylaxis, radiological studies including penile Doppler ultrasound, CT and MRI may be helpful in directing early surgical intervention due to the advanced vascular compromise and extent of necrosis.

**Conflicts of interest**

None.

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