Penile Calciphylaxis
Crystal Rathore, MD, Arya Loghmani, DO, Josh Larrazolo, MD, Jessica Bordes, MD, Jack Harbert, MD, Seema Walvekar, MD, Jonathan Somma, MD
Department of Medicine, LSU Health Sciences Center, New Orleans, LA

Introduction: Calciphylaxis, or calcific uremic arteriolopathy, is a rare skin disorder that presents as intensely painful areas of skin ischemia and necrosis and carries a high morbidity and mortality rate. The skin lesions of calciphylaxis normally involve adipose-rich areas such as abdomen and thighs, though in much rarer cases have involved the penis. Calciphylaxis is most commonly associated with end-stage renal disease (ESRD). Other associated risk factors include hyperphosphatemia, hypercoagulable states, long-term dialysis use, and warfarin use. Diagnosis requires a skin biopsy showing intravascular calcification and thrombosis. Treatment is not definitive and therefore necessitates a multimodal approach including a sodium thiosulfate trial, addition of phosphate binders, thorough wound care, and aggressive pain management. In the case of penile calciphylaxis, strong consideration is given to penectomy. Despite a collaborative effort to mitigate risk factors, calciphylaxis, and in particular penile calciphylaxis, is a highly lethal disorder with a survival of only several months.

Case: A 50-year-old man with past medical history of ESRD on HD and mechanical mitral valve replacement on warfarin presented with one month of progressive penile pain and swelling, which worsened to an ulcerated lesion at the glans penis and phimosis. CT scan of the pelvis demonstrated marked diffuse atherosclerotic disease with wall calcification of all major vessels, as well as questionable mild soft tissue edema and skin thickening of the penis. Infectious workup was negative for chlamydia/gonorrhea, HIV, HSV, and syphilis. The patient was evaluated by urology and taken to the operating room for circumcision and penile biopsy given concern for malignancy. Frozen section for cancer was negative. Final surgical pathology results revealed necrotic ulcer with thrombosis and intravascular calcification at the ulcer base. Dermatology was consulted for review of biopsy. Findings were deemed most consistent with calciphylaxis, and treatment plan was coordinated with nephrology to begin sodium thiosulfate with dialysis. The patient continued to complain of intense pain despite scheduled and prn oral and IV pain medications. Palliative care and wound care were consulted for symptom management. The patient was offered penectomy given refractory pain despite medication adjustment, however he declined. And although it is recommended to discontinue warfarin in the setting of calciphylaxis as it can worsen skin necrosis, the decision was made to continue warfarin given presence of his mechanical valve and otherwise limited options due to ESRD.

Discussion: Penile calciphylaxis is particularly rare because of the rich vascular network in the area. However, if extensive enough, it can be devastating. Microvascular calcification leads to a cascade of vascular endothelial injury and consequent narrowing and thrombosis of the vessels, which ultimately leads to tissue necrosis from reduced blood flow. There is no approved treatment for calciphylaxis. A collaborative approach involving nephrology, dermatology, wound care, and palliative care is necessary to try and reduce risk factors. A trial of sodium thiosulfate is suggested but with uncertain efficacy and make take weeks to months to observe clinical response. In treatment resistant penile calciphylaxis, penectomy is warranted to
potentially prevent months of pain and poor quality of life. Still, in spite of maximal efforts to contain this rare skin disorder, the prognosis of calciphylaxis is poor and mortality risk remains very high.