Penile Pain in a Hemodialysis Patient

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Case Description

A 50-year-old man with a history of diabetes mellitus and ESKD on hemodialysis for 3 years presented with worsening discomfort in the genital region. He was afebrile and the physical examination demonstrated a circumcised phallus with a black necrotic area on the glans (Figure 1) with stenotic meatus. There was tenderness over the glans but no urethral discharge or ulcerations. He was almost anuric at baseline.

Laboratory data were significant for a serum creatinine of 5.24 mg/dl, erythrocyte sedimentation rate of 120 mm/h (2–37), parathyroid hormone level of 81 pg/ml (<300 pg/ml for ESKD), calcium of 8.2 mg/dl (8.2–10.3), phosphorus of 3.4 mg/dl (2.4–4.6) and a serum albumin of 2.6 g/dl (3.2–5). He did have uncontrolled hyperphosphatemia and secondary hyperparathyroidism in the past.

Computed tomography scanning demonstrated diffuse vascular calcifications throughout the abdomen and pelvis (Figure 2). Review of previous lower extremity angiogram revealed a heavily calcified internal pudendal artery with >90% stenosis throughout, as well as opacification of the penile vessels. We diagnosed the patient with penile calcific uremic arteriolopathy (CUA) and started sodium thiosulfate therapy. Unfortunately, the patient died, likely secondary to acute myocardial infarction, a few weeks after discharge from the hospital at a long-term care facility.

CUA, also known as calciphylaxis, is characterized by calcification of arterioles and capillaries in the dermis and subcutaneous tissue, resulting in skin necrosis. It is associated with high morbidity and mortality, largely secondary to sepsis. Although most lesions occur on the lower extremities and abdomen, the penis is a rare site of involvement (1). Hyperphosphatemia, hypercalcemia, uncontrolled secondary hyperparathyroidism, hypoalbuminemia, diabetes, warfarin use, and protein C/S deficiency are some of the risk factors for CUA, and skin biopsy is considered the gold standard for diagnosis. Typical histologic findings include dermo-hypodermal and pannicular arteriolar calcification, microthrombosis, and fibrointimal hyperplasia leading to ischemia and intense septal panniculitis (2). However, penile CUA is essentially a clinico-radiologic diagnosis because biopsy might lead to progression of necrosis. Demonstration of vascular calcifications in the pelvic region, particularly pudendal vessels, strongly suggests the diagnosis (3). Once diagnosed, treatment is multi-interventional with a focus on wound and pain management, management of bone mineral disease, ensuring dialysis adequacy, sodium thiosulfate, and management of other risk factors (e.g., discontinuation of warfarin). Retrospective data suggest that sodium thiosulfate is effective in promoting wound healing but reports of a survival benefit are inconsistent (4,5). In fact, the 1-year mortality remains >50% in patients with CUA despite treatment (2).

Teaching Points

- Nephrologists should have a high index of suspicion for CUA when patients on dialysis complain of pain or discomfort in the genitalia.
- Penile calciphylaxis is primarily a clinico-radiologic diagnosis.
- This condition is associated with high mortality and sodium thiosulfate therapy must be considered when possible despite equivocal evidence at this time.

Disclosures

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Author Contributions

C. Cabrera Fermin wrote the original draft, A. Koratala provided supervision, and S. Singh reviewed and edited the manuscript.

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