Ulceration and Necrosis of Fingers and Toes in an ESKD Patient

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Case Description
A 54-year-old gentleman presented with necrotic cutaneous areas on his toes and fingers after failure of his kidney transplant requiring a return to hemodialfiltration after 15 years of adequate allograft function. He had a background history of tertiary hyperparathyroidism, rheumatoid arthritis, Addison’s disease, and recurrent pulmonary emboli. X-rays were performed of his affected hand and foot (Figures 1 and 2). The patient required amputation of multiple digits due to ischemia caused by calcific uremic arteriolopathy (CUA). In addition to parathyroidectomy, he commenced intravenous sodium thiosulfate after dialysis, stopped warfarin, and had an intensification of his hemodialfiltration adequacy, with increased control of his serum calcium and phosphate (1). At the time of writing, the patient’s CUA had not progressed extensively since diagnosis in 2013, and he remains on intermittent hemodialfiltration.

Discussion
CUA is a rare but serious disorder that presents with skin ischemia and necrosis, which is often painful (2). It most commonly occurs in patients who have ESKD and are on maintenance dialysis; however, it may also occur in patients who have undergone a kidney transplant and patients with CKD. It carries a significant morbidity and mortality, and is an important clinical diagnosis to consider in patients with CKD.

CUA typically presents with areas of excruciatingly painful ischemic necrosis. These lesions typically develop in areas with the greatest adiposity and most commonly involve distal lower extremities, proximal lower extremities, and trunk and distal upper extremities (2). Early lesions may present as painful plaques or subcutaneous nodules that progress to ischemic ulcers with eschars.

The diagnosis of CUA is on the basis of focused physical examination and awareness of additional clinical features (warfarin use, obesity, hyperphosphatemia, elevated parathyroid hormone trend over the preceding months) (3). A dermatologist review should be requested for initial evaluation to help eliminate mimics of CUA and, although radiologic investigations may be suggestive of the diagnosis, a skin biopsy may be required in some patients. The differential diagnosis for CUA includes but is not limited to atherosclerosis, cholesterol embolization, warfarin necrosis, vasculitis, and purpura fulminans.

There are no approved pharmacological treatments for CUA, although multiple medications are in use off-label. A multidisciplinary approach should be adapted to the initial management of any patient with CUA, including wound care specialists, nephrologists, pain care specialists, and dermatologists. Wounds with a heavy necrotic burden are at high risk for infection and may require plastic surgery specialty...
input for consideration of debridement/amputation where appropriate.

Because CUA is a rare disorder, high-quality randomized evidence is lacking; however, a number of strategies are usually used on the basis of the best available evidence to date:

- Patients with hyperphosphatemia are treated with a noncalcium-containing phosphate binder to decrease the rate of vascular calcification.
- In patients with an elevated serum parathyroid hormone parathyroidectomy may be required, cinacalcet therapy may be initiated where parathyroidectomy is not thought to be appropriate. There is also evidence from the EVOLVE study (4) that cinacalcet may be protective against the development of CUA.
- A trial of sodium thiosulfate should be considered for at least 3 to 4 weeks of therapy. Although this therapy is off label, it is thought to have vasodilatory and antioxidant properties, and is usually given after each dialysis session (5).

Dialysis prescriptions should also be optimized to enhance clearance of phosphate and avoidance of excessive calcium administration (stopping vitamin D/calcium supplements), in addition to considering cessation of warfarin where appropriate.

Teaching points

- CUA should be considered in patients with ESKD who present with areas of ulceration that are concerning for necrosis.
- In select patients with ESKD presenting with CUA, treatment options may include: a noncalcium-containing phosphate binder, cinacalcet therapy or consideration for a parathyroidectomy, and a trial of intravenous sodium thiosulfate.
- Surgical resection and/or amputation may be considered if the ischemic burden of the affected area is significant.

Disclosures

D. Sexton reports being a shareholder in patientMpower. All remaining authors have nothing to disclose.

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Author Contributions

R. Piggott wrote the original draft; and G. Mellotte and D.J. Sexton reviewed and edited the manuscript.

References


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Figure 2. | Plain (oblique and lateral) x-rays of the patient’s affected hand. (A) and (B) show severe extensive diffuse vascular calcification in medium-sized and smaller arterial blood vessels characteristic of calcific uremic arteriolopathy.