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Classic Case

Calcific uraemic arteriolopathy: A rare but devastating complication of end-stage renal failure

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Abstract

Calcific uraemic arteriolopathy is a rare complication of end-stage renal failure. It carries a grave prognosis with 1-year survival of under 50%. It occurs due to subcutaneous small vessel calcification, thrombosis, with subsequent tissue necrosis.

We described a case of calcific uraemic arteriolopathy in a 58-year-old man who presented with violaceous indurations over bilateral lower limbs, as well as large necrotic ulcer with adjacent eschars at the lower abdomen. Although skin biopsy is the gold standard for diagnosis, it is often avoided due to potential poor wound healing. On the other hand, in radiographs or computed tomography, fine linear or serpiginous subcutaneous calcifications are typical manifestations, which represent underlying small vessel calcifications. Radiological examinations, therefore, play an important role to establish the diagnosis.

Keywords: Calcific uraemic arteriolopathy, Calciphylaxis, Renal failure.

Background

Subcutaneous calcifications may often be dismissed, especially if there is significant pathology in the visceral organs or the skeletal structures. This article demonstrates a classic case of calcific uraemic arteriolopathy, a rare but serious complication of end-stage renal failure, which manifests as subcutaneous calcifications.

Case summary

A 58-year-old man was delivered to the emergency department in May 2023 with bilateral lower limb pain. He had morbid obesity, diabetes mellitus, and end-stage renal failure on peritoneal dialysis. Physical examination showed violaceous and plaque-like indurations over bilateral lower limbs, without open wound or signs of infection. Radiographs of bilateral lower limbs showed extensive subcutaneous fine linear calcific densities (Figure 1). He was discharged after the pain subsided with non-opioid analgesics.

In November 2023, he was admitted to the renal ward for blocked Tenckhoff catheter. A large necrotic ulcer with adjacent eschar was discovered at the left lower abdomen (Figure 2). The patient reported progressive pain and pruritis from his lower abdomen to bilateral lower limbs in the past month. He developed fever during his hospital stay, and computed tomography of the abdomen and pelvis was performed, showing subcutaneous small arteriole calcifications of the lower abdomen (Figure 3). While some calcified plaques were noted along the aorta, there were no extensive calcifications of the visceral organs, such as the kidneys, adrenals or pancreas (Figure 4). Clinical and radiological features were suggestive of calcific uraemic arteriolopathy. Wound swab showed multi-drug resistant Acinetobacter baumannii, and intravenous cefoperazone / sulbactam, metronidazole and minocycline were prescribed. Wound debridement was performed, and a histopathological examination of the excised skin tissue showed necrosis in the dermis and subcutis, as well as small vessel wall calcifications and thrombi, which were consistent with calcific uraemic arteriolopathy (Figure 5). Unfortunately, the patient deteriorated clinically, and succumbed due to multi-organ failure in one month.

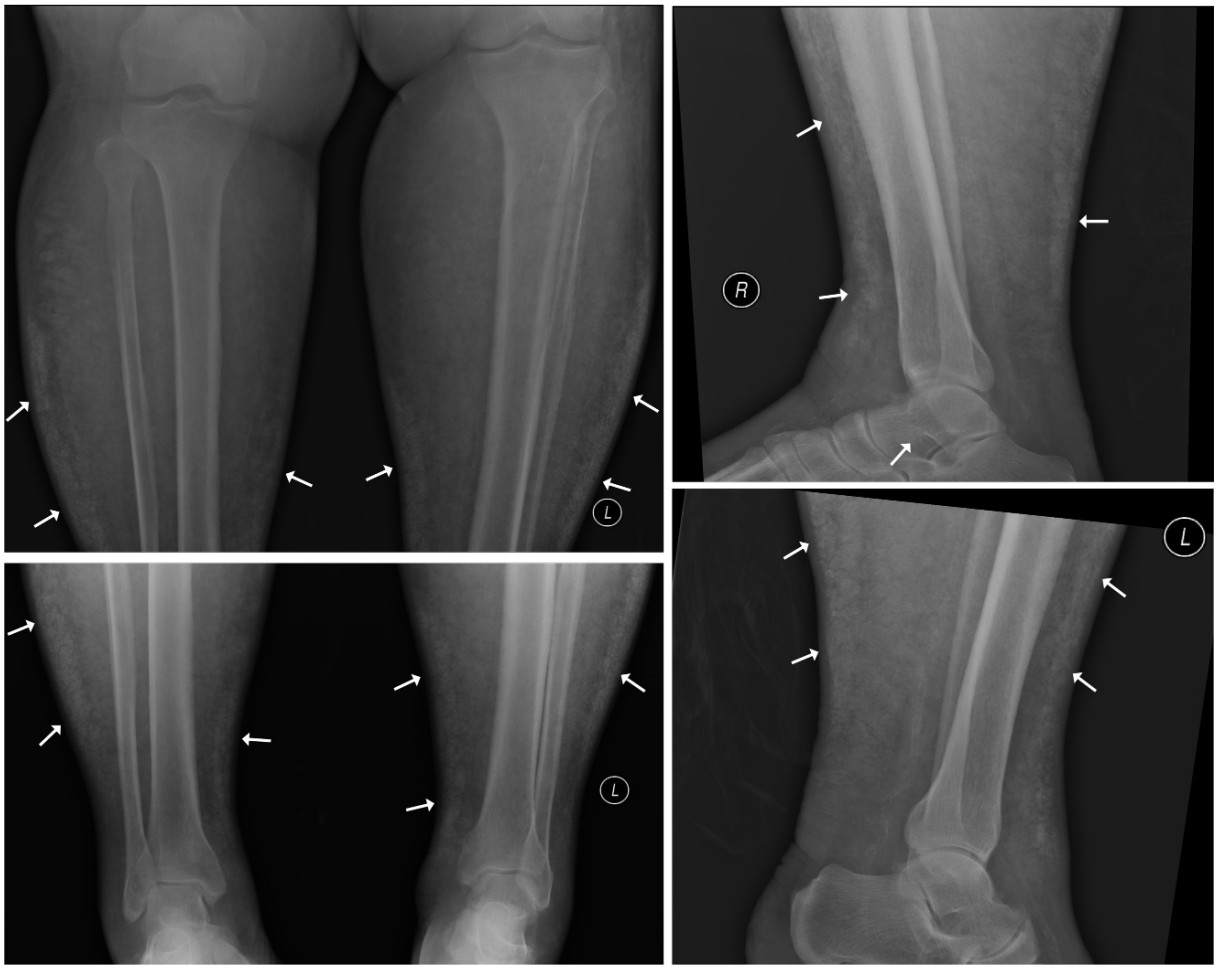

Figure 1. *Radiographs of the patient's bilateral lower limbs show extensive subcutaneous fine linear calcific densities (white arrows), suggestive of calcific uraemic arteriolopathy.*

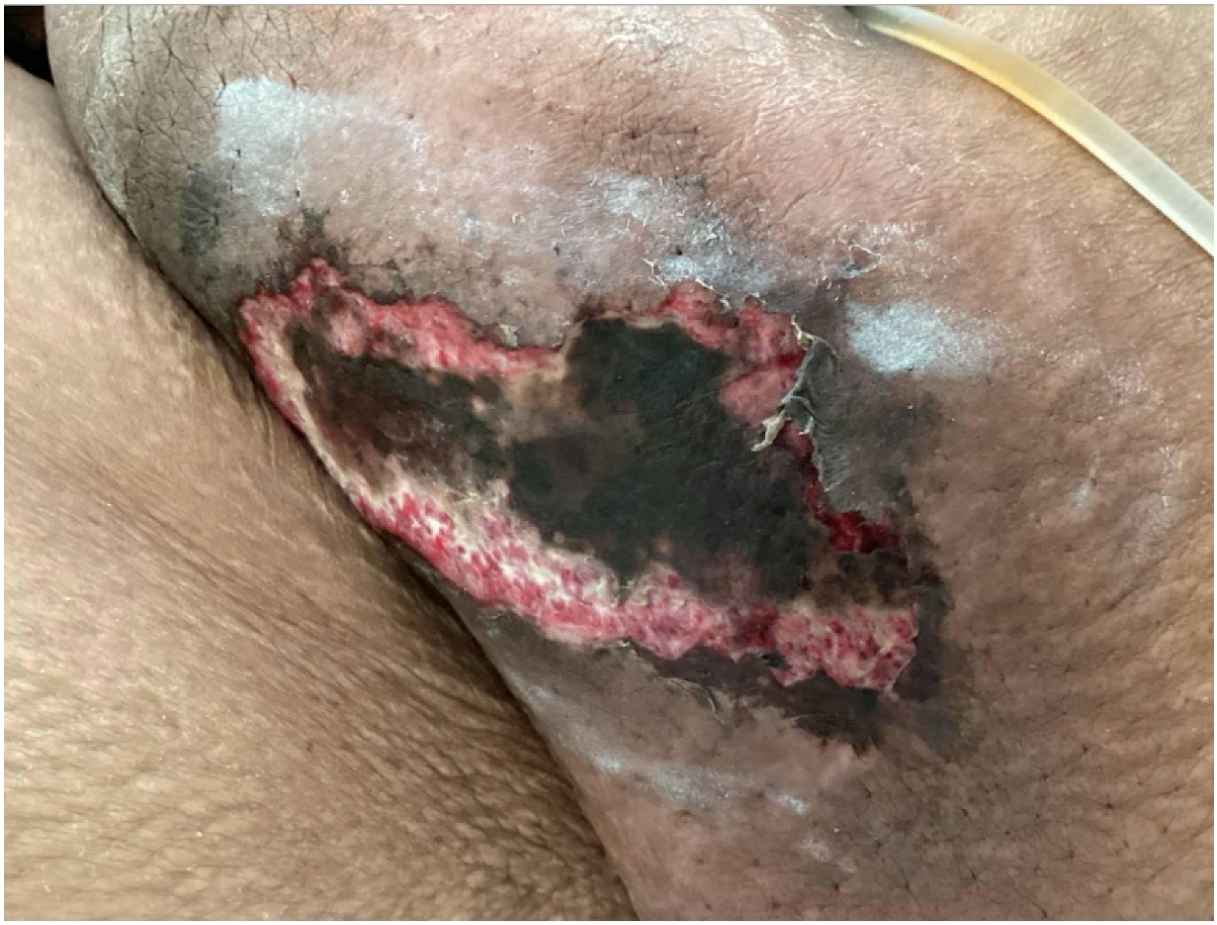

Figure 2. *Image of the patient's left lower abdomen lateral and caudal to the Tenckhoff catheter insertion site shows a large 20cm x 15cm necrotic ulcer with adjacent eschar.*

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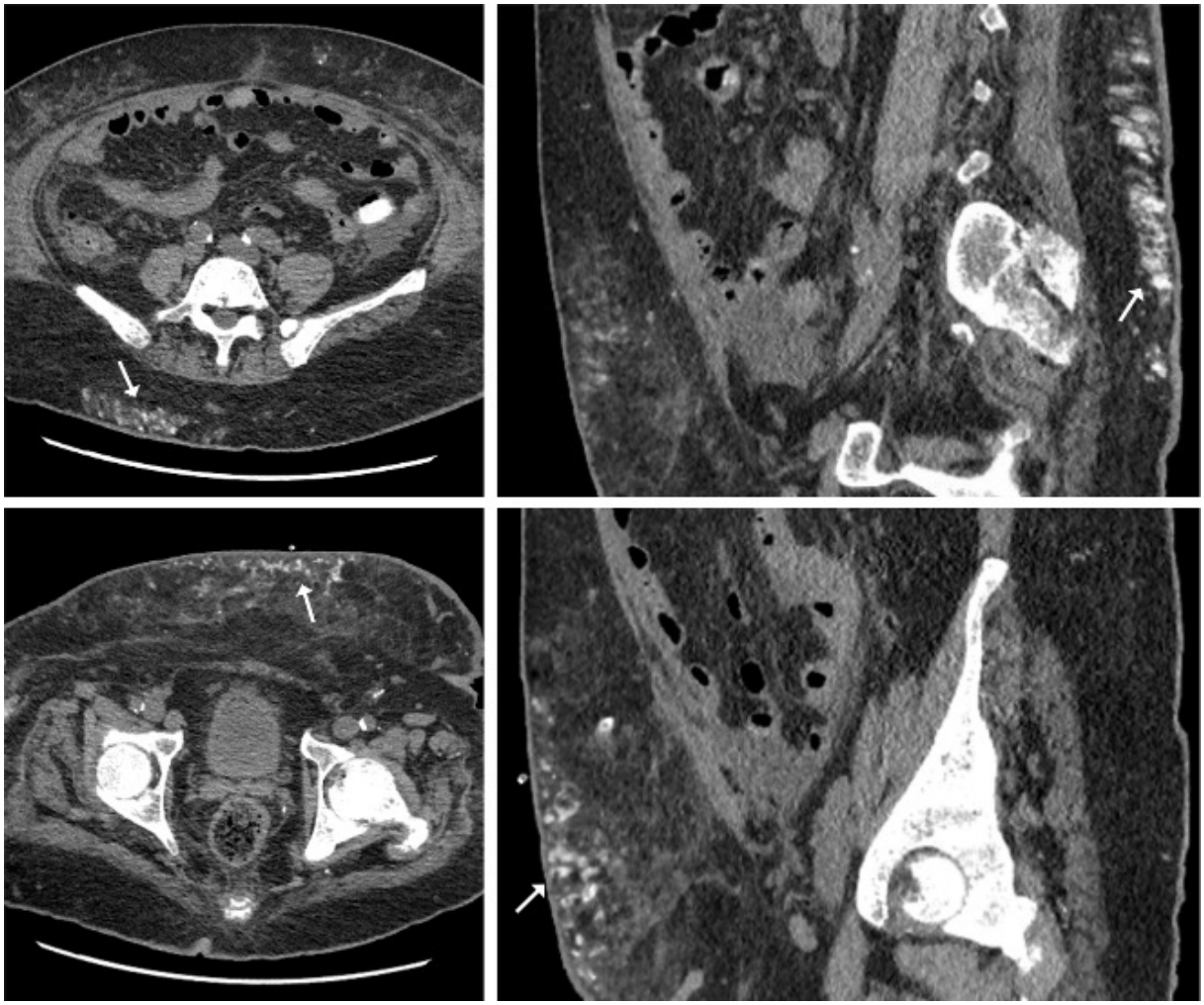

Figure 3. *Non-contrast computed tomography of the patient's abdomen and pelvis shows subcutaneous small arteriole calcifications of the lower abdomen (white arrows), suggestive of calcific uraemic arteriolopathy.*

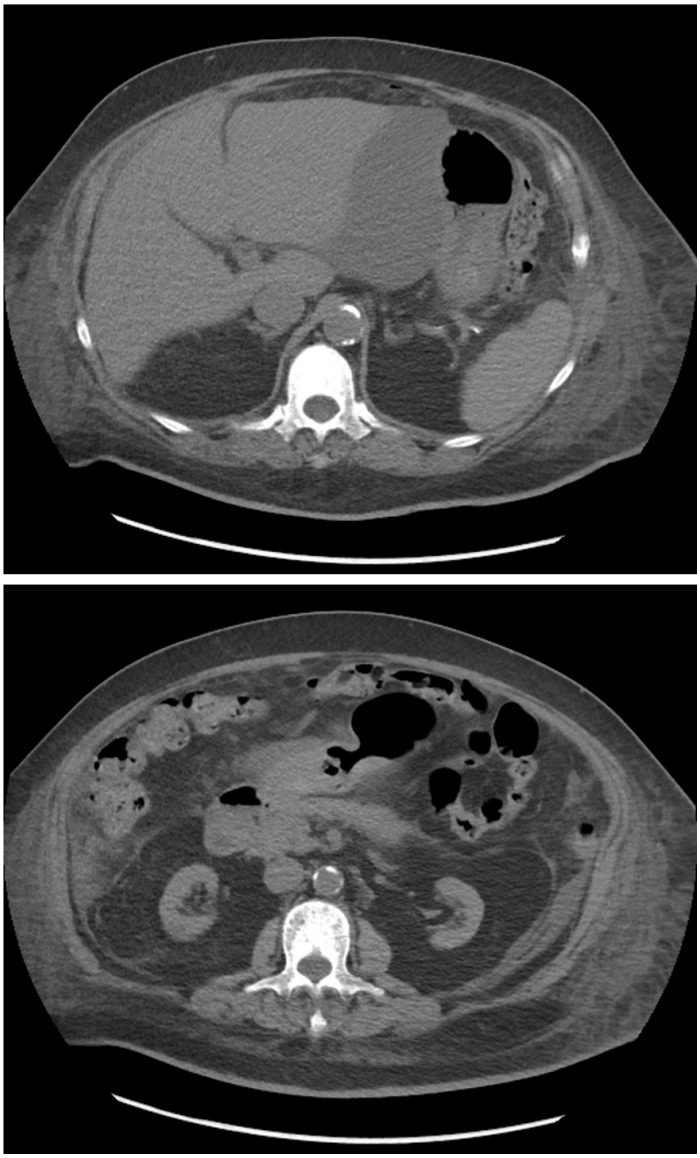

Figure 4. *Non-contrast computed tomography of the patient's abdomen and pelvis shows some calcified plaques along the aorta, though there are no extensive calcifications of the visceral organs, such as the kidneys, adrenals or pancreas.*

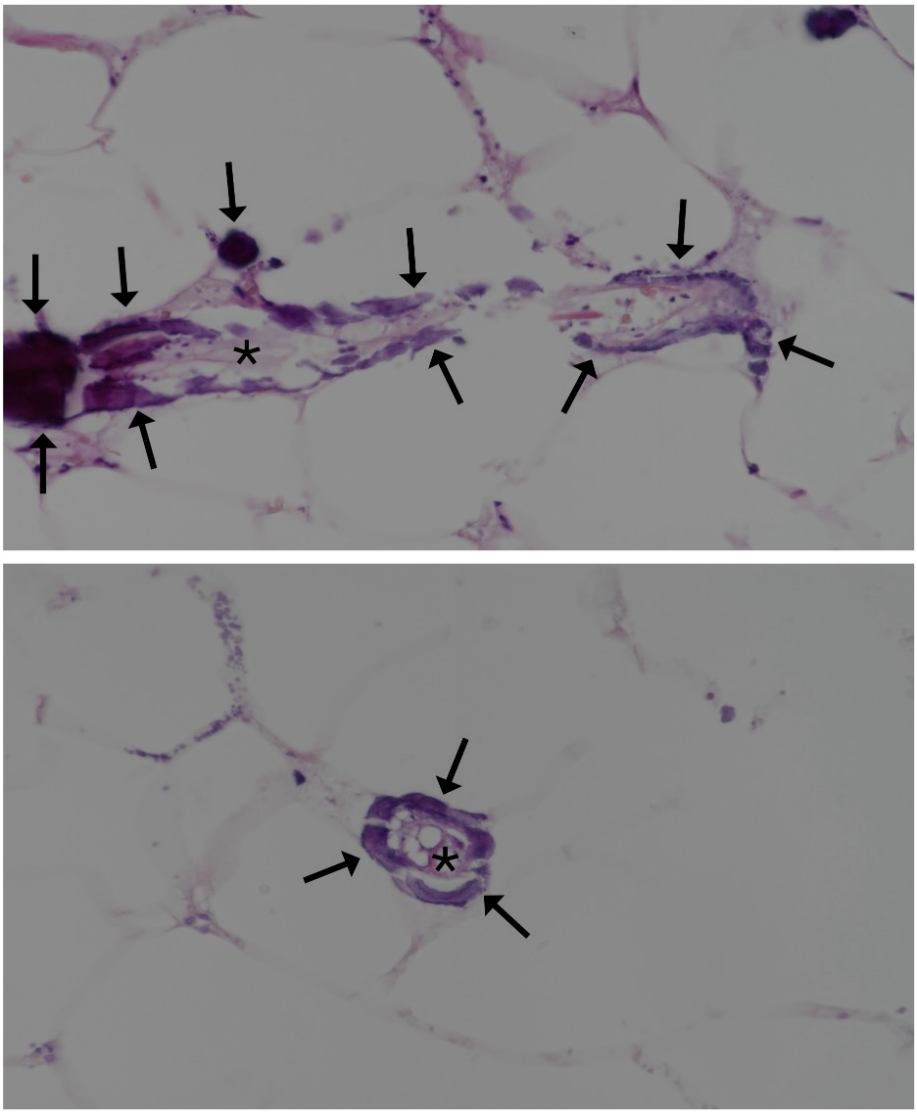

Figure 5. *Micrographs of the specimen (haematoxylin and eosin stain, 400x magnification) from debrided skin tissue from the patient's left lower abdominal wound show small vessel wall calcifications (black arrows) and thrombi (black asterisks), consistent with calcific uraemic arteriolopathy.*

Discussion

Calcific uraemic arteriolopathy, also known as calciphylaxis, is a rare complication of end-stage renal failure, occurring in less than 5% of patient on dialysis [1]. It carries a grave prognosis with 1-year survival of under 50% [2]. Its pathogenesis has yet been entirely elucidated, although hyperparathyroidism, deranged calcium and vitamin D levels, as well as chronic inflammation are postulated to play key roles. Risk factors include obesity, diabetes and female sex, the first two of which are present in this patient [2-3].

Calcific uraemic arteriolopathy typically presents with intense pain, pruritus and subcutaneous induration in the regions of greatest adiposity, more commonly in the lower limbs and the abdomen. It initially appears violaceous, which may eventually progress to eschars, and non-healing ulcers with cutaneous necrosis [1]. While it is usually diagnosed clinically, skin biopsy remains the gold standard. Histopathological examination usually shows small vessel calcification, endovascular fibrosis, thrombosis, as well as subcutaneous tissue necrosis [1-2]. Yet, skin biopsy is often avoided due to potential poor wound healing, especially when there is a superimposed infection. Radiological examinations, as illustrated in this case, are helpful to establish the diagnosis. In plain radiographs, fine linear subcutaneous calcifications are seen, which represent underlying small vessel calcifications. They are usually present in the lower limbs and the abdomen, corresponding to the symptomatic regions with skin and subcutaneous changes. Of note, this contrasts with tumoral calcinosis, another uncommon manifestation associated with chronic renal failure, which shows lobular calcified masses at periarticular locations with calcium sedimentation. Computed tomography can better delineate the distribution of calcification in small vessels, as well as exclude any presence of deep-seated infection [4-5].

There is no established treatment for calcific uraemic arteriolopathy in current literature. In view of a high infection risk, meticulous wound care and debridement of necrotic tissue are essential [2-4].

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To conclude, calcific uraemic arteriolopathy is a rare but devastating complication in patients with end-stage renal failure. It has distinctive radiological features which can help establish the diagnosis.

Ethics approval

The patient was treated in accordance with the Declaration of Helsinki. He provided informed consent for all treatments and procedures. The patient's next-of-kin provided informed consent for publication.

Conflicts of interest and source of funding

All authors declared no conflicts of interest. This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

References

1. Colboc H, Moguelet P, Bazin D, Carvalho P, Dillies AS, Chaby G, et al. Localization, morphologic features, and chemical composition of calciphylaxis-related skin deposits in patients with calcific uremic arteriolopathy. JAMA Dermatol 2019;155:789-96. doi: 10.1001/jamadermatol.2019.0381.

2. Weenig RH, Sewell LD, Davis MD, McCarthy JT, Pittelkow MR. Calciphylaxis: natural history, risk factor analysis, and outcome. J Am Acad Dermatol 2007; 56:569-79. doi: 10.1016/j.jaad.2006.08.065.

3. Jovanovich A, Chonchol M. Calcific uremic arteriolopathy revisited. J Am Soc Nephrol 2016;27:3233-5. doi: 10.1681/ASN.2016040480.

4. Jeong HS, Dominguez AR. Calciphylaxis: controversies in pathogenesis, diagnosis and treatment. Am J Med Sci 2016;351:217-27. doi: 10.1016/j.amjms. 2015.11.015.

5. Bleibel W, Hazar B, Herman R. A case report comparing various radiological tests in the diagnosis of calcific uremic arteriolopathy. Am J Kidney Dis 2006; 48:659-61. doi: 10.1053/j.ajkd.2006.05.031.