Acral calciphylaxis: distinguishing between a rare entity and peripheral vascular disease

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Background

- Calciphylaxis is a life-threatening disease due to progressive subcutaneous and dermal small vessel calcification → thrombosis[1,2]
  - Most common in those with end-stage renal disease (ESRD), especially dialysis patients and renal transplant recipients, though non-nephrogenic variants exist[3-5]
  - Presents with non-inflammatory retiform purpura, ulceration, tissue ischemia, and skin necrosis[1,2]
  - Most commonly affects adipose-rich tissue such as the trunk, buttocks, or pannus[1,2]
- **Acral calciphylaxis** = rare subtype involving digits of extremities and genitalia[2]
- Mortality rates for calciphylaxis: 44-50% within 12 months of diagnosis[6]
- Early dx and management important due to poor prognosis but often do not happen due to limited knowledge, lack of standardized dx criteria, & difficulty in differentiation from peripheral vascular disease (PVD)

Objective

To characterize clinical features and assess mortality outcomes for a series of patients with lesions suspicious for acral calciphylaxis

Methods

Inpatient dermatology consult database was screened for potential cases seen between January 1, 2012, and June 30, 2023, across 2 hospitals

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Patients with potential acral calciphylaxis were IDed using search terms: “acral calciphylaxis,” “acral calci,” “calciphylaxis of digits,” “calciphylaxis of fingers or toes,” “calciphylaxis of hands,” “calciphylaxis of feet,” “calciphylaxis of extremities,” “penile calciphylaxis,” “penile calci,” “calciphylaxis of penis,” and “calciphylaxis of genitalia”

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Each patient’s clinical notes, labs, & imaging were reviewed for coagulopathies, embolic disease, infection, septic vasculitis, and cryoglobulinemia. Patients with any of these conditions which led to their presentation aside from calciphylaxis versus PVD were excluded

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Final sample included 22 patients

Results

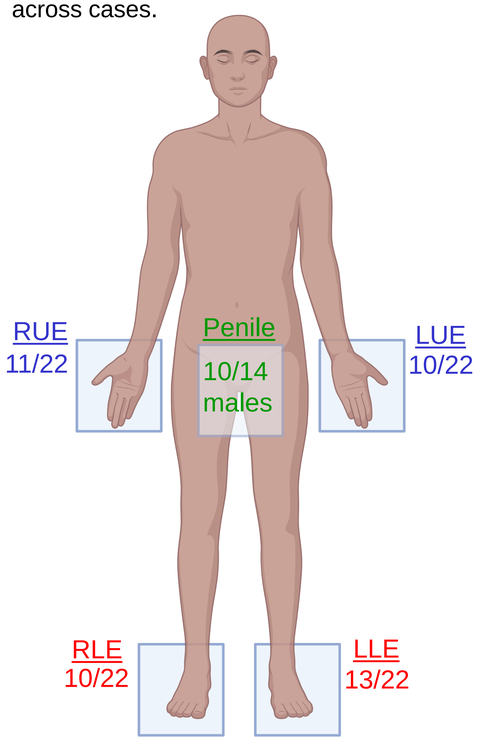
Table 1. Demographic and clinical characteristics of cases.

Characteristic	Total (n = 22)
Demographics	
Age at diagnosis, median years (IQR)	55.5 (49 – 63)
Male, n (%)	14 (63.6%)
Race & Ethnicity, n (%)	
Non-Hispanic Black	7 (31.8%)
Hispanic White	15 (68.2%)
Co-morbidities, n (%)	
ESRD	19 (86.4%)
T2DM	18 (81.8%)
Underlying PVD	18 (81.8%)

n, number; IQR, interquartile range; ESRD, end-stage renal disease; T2DM, type 2 diabetes mellitus; PVD, peripheral vascular disease.

- 5/22 patients had involvement of both upper and lower extremities
- 15/22 patients had involvement of contralateral extremity
- In 12/22 patients, vascular surgery could not exclude PVD as the etiology of ≥1 of the patient’s ischemic lesions, including 6/12 with steal syndrome secondary to the hemodialysis (HD) arteriovenous (AV) fistula on their ipsilateral extremity
- 5/22 patients had subsequent amputations of affected extremity
- 7/22 died within 12 months of admission. Of the 10 confirmed dead at time of analysis, median survival was 156.5 days
- 2/10 deaths related to calciphylaxis

Figure 1. Anatomical distribution of lesions across cases.



Conclusions

- Most patients were Hispanic White or non-Hispanic Black with ESRD, T2DM, & PVD
- For large % patients, PVD contributes to acral lesion development and possible vascular intervention may be required
- Dermatologists should consider steal syndrome in their ddx for patients with ischemic acral lesions who have HD AV fistulas in ipsilateral limbs
- Dermatology & vascular surgery collaboration is needed to develop clinical criteria for the diagnosis of acral calciphylaxis and to improve clinical outcomes

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