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

Background: Calciphylaxis is a life-threatening disease due to progressive subcutaneous and dermal small vessel calcification resulting in thrombosis. Acral calciphylaxis is a rare subtype causing digital ischemia. There are no standardized criteria for its diagnosis, and distinguishing from peripheral vascular disease (PVD) is difficult. This study aimed to capture clinical characteristics and mortality outcomes for a series of patients with lesions suspicious for acral calciphylaxis.

Case Presentation: An inpatient dermatology consult database containing patients seen across 2 hospitals over 12 years was screened for potential cases. Those with other causes aside from calciphylaxis versus PVD were excluded, leaving a final sample of 22 patients. A retrospective analysis was conducted, and mortality outcomes were determined.

Results: Mean age was 55.7 years and 65.2% of patients were male. 69.6% were White-Hispanic, and 30.4% were Black. Mean body mass index was 26.2 kg/m². Patients had the following comorbidities: 100% had chronic kidney disease with 86.4% also exhibiting end-stage renal disease (ESRD), 81.8% had type 2 diabetes mellitus (T2DM), and 81.8% had underlying PVD. Six patients had steal syndrome. The median number of lesions was 3, and among those with both acral and non-acral lesions, 60% had the acral lesion develop first. Twelve patients (54.5%) had lesions on an upper extremity, 15 (68.2%) had lesions on a lower extremity, and 5 (22.7%) had lesions on both an upper and lower extremity. Fifteen patients (68.2%) had symmetric lesions. Based on imaging and ankle-brachial indexes, vascular surgery surmised that 50% of acral lesions were likely due to PVD. Four patients had to undergo amputations of extremities with lesions. Eight patients (36.4%) died within 12 months of admission. Of the 9 patients (40.9%) who had expired at time of analysis, median survival was 75.5 days. Two deaths were determined to be directly due to calciphylaxis.

Conclusion: Most patients presenting with acral lesions had underlying ESRD and PVD. A large proportion of acral lesions may be due to PVD rather than acral calciphylaxis. High morbidity and mortality necessitate dermatology and vascular surgery collaboration to further characterize the patterns of clinical presentation and to standardize diagnostic criteria.