Clin Kidney J (2013) 6: 653–654
doi: 10.1093/ckj/sft110
Advance Access publication 20 September 2013

CKJ

Images in Nephrology
(Section Editor: G. H. Neild)

Calciphylaxis

Ali Nayer[1], Loay Salman[1] and Arif Asif[2]

[1]Division of Nephrology and Hypertension, University of Miami, Miami, FL, USA and [2]Division of Nephrology and Hypertension, Albany Medical College, Albany, NY, USA

Correspondence and offprint requests to: Ali Nayer; E-mail: anayer@med.miami.edu

Keywords: calcific uremic arteriolopathy; calciphylaxis

A 49-year-old man presented with dry gangrene of the fingers and penis. The past medical history was notable for type 1 diabetes mellitus since age 13, hypertension, dyslipidemia, coronary artery disease, peripheral vascular

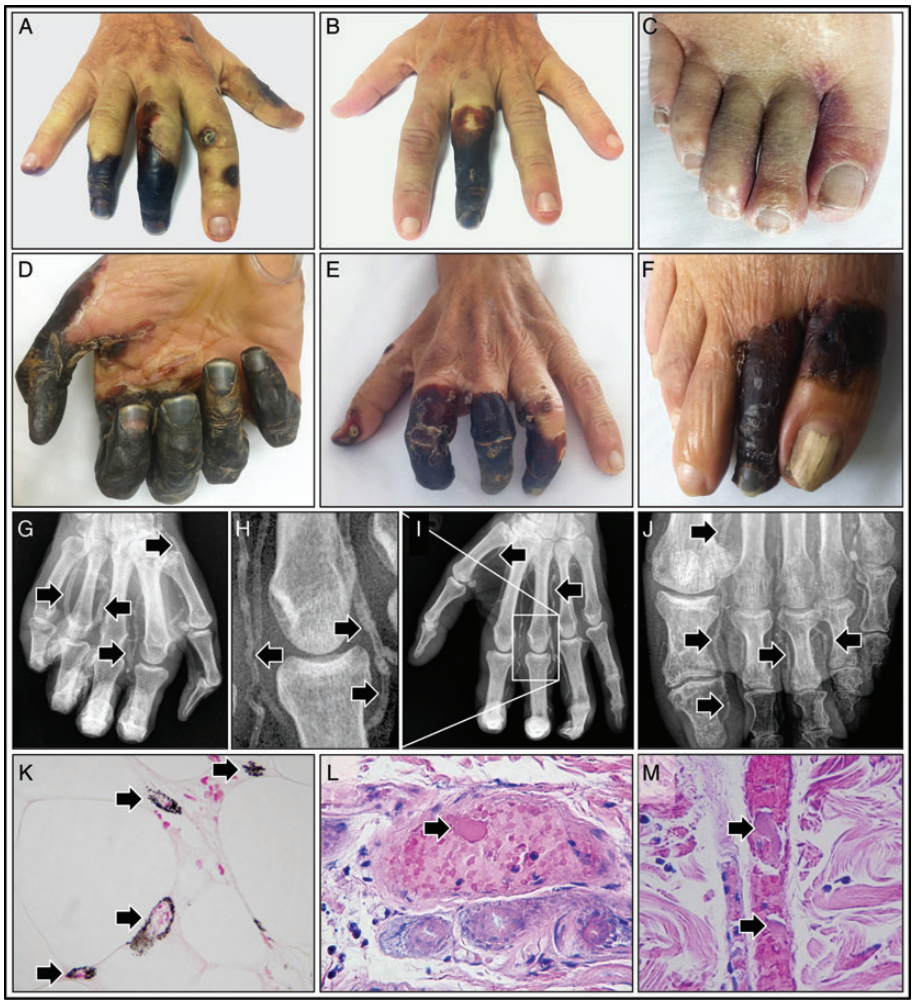

Fig. 1. Dry gangrene of the fingers (A and B) and dusky red discoloration of the right great toe (C) are shown. Progression of the gangrene of the fingers (D and E) and development of gangrenous necrosis of the toes (F) are shown. Plain radiographs demonstrating marked vascular calcification of the hands and feet (G–J). Skin biopsy revealing calcification (K) and microthrombosis (L and M) of small subcutaneous blood vessels consistent with calciphylaxis. Tissue sections were stained according to von Kossa method (K) and with Hemotoxylin and eosin stain (L and M).

disease and end-stage renal disease on hemodialysis for 5 years. Physical examination revealed dry gangrene of the fingers (Figure 1A and B), penis and scrotum. Dusky red discoloration of the right lateral great toe was also noted (Figure 1C). Laboratory tests demonstrated hypoalbuminemia, hyperphosphatemia and mild secondary hyperparathyroidism. Cryoglobulins, cold agglutinins, cryofibrinogen, lupus anticoagulant and antibodies directed against cardiolipin, β2 glycoprotein I, myeloperoxidase, proteinase 3, platelet factor 4, nuclear antigens, hepatitis B and C viruses were not detected. Serum concentrations of antithrombin, protein S and protein C were normal. Prothrombin G20210A mutation was not detected. Echocardiography demonstrated preserved ejection fraction and no atrial septal defect. While the diagnostic workup was in progress, gangrene of the fingers and toes worsened (Figure 1D–F). In addition, several bullae appeared on brown to purple linear bands on the lateral thighs. Radiographs of hands and feet revealed marked vascular calcification (Figure 1G–J). A skin biopsy revealed calcified subcutaneous small blood vessels consistent with calciphylaxis (von Kossa stain) (Figure 1K). In addition, microthrombi were noted in subcutaneous small blood vessels (Figure 1L and M). There was no evidence of cholesterol emboli.

Calciphylaxis, also known as calcific uremic arteriolopathy, is characterized by slowly progressive skin necrosis accompanied by calcification and thrombosis of small- and medium-sized arteries [1–3]. It affects mainly middle-aged and older individuals in the setting of renal disease. Deranged calcium and phosphate metabolism is considered the pivotal pathogenesis of calciphylaxis. Radiographs of the affected areas may reveal vascular calcification. Histologically, small- and medium-sized arteries demonstrate medial calcification, intimal hyperplasia and thrombosis. The mortality approaches 80% in patients with renal disease. The quest for an effective treatment continues.

Author contributions

AN, LS, and AA prepared the figure and wrote the manuscript.

Conflict of interest statement. The results presented in this paper have not been published previously in whole or part, except in abstract form.

References

1. Wilmer WA, Magro CM. Calciphylaxis: emerging concepts in prevention, diagnosis, and treatment. *Semin Dial* 2002; 15: 172–186
2. Weenig RH. Pathogenesis of calciphylaxis: Hans Selye to nuclear factor kappa-B. *J Am Acad Dermatol* 2008; 58: 458–471
3. Vedvyas C, Winterfield LS, Vleugels RA. Calciphylaxis: a systematic review of existing and emerging therapies. *J Am Acad Dermatol* 2012; 67: e253–e260

Received for publication: 3.8.13; Accepted in revised form: 6.8.13