Non-Uremic Calciphylaxis

James Connors1, DPM, Lauren Kishman1, DPM, Joris Claessens1, DPM, Stella Chundu DPM2

1. Podiatric Medicine and Surgery Resident, HealthSpan/Cleveland Clinic
2. Faculty of the Podiatry Residency Training program at the Cleveland Clinic and Associate of the American College of Foot and Ankle Surgeons

Pre-tx:

10 months post-tx:

Discussion

Treatment of calciphylaxis requires control of the calcium-parathyroid hormone axis [8-10]. Sodium thiosulfate is a chelating agent with anti-oxidant efficacy that is utilized in calcifying nephro lithiasis and tumor-induced calcinosis [8]. Sodium thiosulfate increases calcium solubility in vascular wall deposits and improves hemodynamics clearance. Anticoagulants inhibit soft tissue calcification by binding to calcified vascular smooth muscle cells, thereby inhibiting progression of the process [9-10]. Prevention of systemic infection is vital. D irected wound care and avoidance of trauma are imperative to successful treatment. There is debate as to whether or not aggressive wound debri deding is warranted in calciphylaxis due to possible re-aggregation or creation of necrotic u lcers. Advocated of aggressive debri deding site reduces wound infection rates thus decreasing the likelihood of sepsis and organ failure. Routine debri deding of bacterial and necrotic eschar is necessary. Surgical debri deding can be combined with split-sk in mesh-graft transplantation. Negative pressure wound therapy may be used adjunctively to increase sk in graft survival in chronic leg ulcers. R evitalization can further enhance the limb salvage rate [7].

Acknowledgements

Trent Marburger MD pathologist responsible for providing the pictures of the histological slides and biopsy description.

References


Case Report

We report on a 55 year old morbidly obese, non-diabetic female admitted for digital gangrene of fingers and toes. Past medical history significant for rheumatoid arthritis (RA) with long-term dextran therapy, non-uremic dialysis patient, non-uremic hyperparathyroid, cardiovascular CVA, and peripheral neuropathy of bilateral hands and feet. Patient initially described the development of blue discoloration of fingers and toes over a period of two weeks which rapidly progressed into necrosis. Coinciding with the development of gangrenous digits was the development of ulcerations to the lower extremities, including a large wound on the posterior right calf and several over the dorsum of bilateral feet. Patient denied prior episodes of digital discoloration with or without cold exposure or Raynaud’s phenomenon. Electrocardiogram revealed a monomorphic tachycardia with evidence of right axis deviation and frequent premature ventricular contractions. On physical examination, pedal pulses were non-palpable but audible. Duplex ultrasonography revealed normal waveforms at the common femoral vein, femoral vein, profunda femoris veins, and common iliac veins bilaterally. No erosive changes or periosteal reactions noted. The remainder of the neurovascular and musculoskeletal examination were within normal limits, including normal waveforms of the ankle and transmetatarsal level bilateral. Severe delayed waveforms at the digital level bilateral.

Pre-tx:

Calcium and phosphorous levels within normal limits. CRP: 0.4 mg/dL. WBC: 11.8 (H). ESR: 0 (H). CRP: 1.2 mg/dL. WBC: 12.8 (H). ESR: 2 (H). No abnormalities noted on full bone density.

Histologic results: Consistent with calciphylaxis (Figure 5)

Figure 3. Calcium and phosphorous levels within normal limits.

Figure 4. ESR and CRP levels.

Figure 5. Histopathologic specimens showing rare interfetel calcium deposits within deep dermis and adipose tissue. Focal calcium deposits within a subcutaneous blood vessel. Small, calcified foci are noted on background of fibrosis in the deep subcutaneous adipose tissue.