A Case of Extraskeletal Calcification –
Calciphylaxis

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Background:
• Calciphylaxis is a rare, often fatal disorder characterized by systemic medial calcification of the arterioles leading to ischemia and subcutaneous necrosis.
• Ischemic changes lead to livedoreticularis and violaceous plaque-like lesions progressing to painful necrotic ulcers.
• Almost exclusively seen in ESRD patients
• Increased risk with:
  – Female sex
  – Obesity
  – High Ca and PO4 levels
  – Medications (Warfarin, phosphate binders, high dose vitamin D)

Diagnosis
• No specific diagnostic laboratory tests
• Clinical suspicion is the single most important feature
• Hypercalcemia, hyperphosphatemia, elevations in the Ca x P product, hyperparathyroidism, and exposure to Ca and Vit D products should raise the suspicion
• Biopsy along with clinical features help confirm the diagnosis
• Bone scanning has been recommended as an alternative to biopsy (especially useful when there is concern that a biopsy could lead to ulcer formation).

Treatment
• Mainly supportive
• No controlled prospective studies comparing different strategies
• Correcting PTH
  – cinacalcet or parathyroidectomy in refractory cases
• Normalizing serum Ca & P abnormalities (product < 55)
  – non-calcium containing phosphate binders
• Sodium thiosulfate has shown significant reduction in pain and skin lesions
• Wound care, pain control and avoidance of local tissue trauma
• Future directions – bisphosphonates, prednisone, and hyperbaric oxygen therapy being studied

Laboratory Workup:
- Hct 29.6
- WBC 6.8
- BUN 31
- Creatinine 9.8
- iPTH 63
- Calcium 7.5
- Phosphorous 8.4
- ESR, CRP, ENA and ANA screen unremarkable.

Skin Biopsy:
Intimal calcification in the small and medium sized blood vessels with necrosis of the overlying skin consistent with calciphylaxis.