Renal Disease and Dermatopathology

Paul K. Shitabata, M.D.
Dermatopathology Institute
Ichthyosis
Histopathology

- Compact layer of orthokeratosis
- May have dermal pigment incontinence
- Invisible dermatosis
Perforating Dermatosis of Renal Disease
Histopathology

- Follicular plug
- Collections of neutrophils adjacent to follicle
- Transepidermal elimination
- Deeper sections
- VVG stain may be helpful
Porphyria
DDX-Porphyria

- Cell poor subepidermal blister
- Subepithelial papillae preserved
- Thickened basement membrane
- Perivascular fibrin by DIF
- Caterpillar bodies
Nephrogenic Fibrosing Dermopathy
(NFD)
Clinical Appearance

- Scleroderma-like condition on the extremities
- Indurated and hardened fibrotic plaques
- Extremities and trunk
- Flexion contractures may result
Epidemiology

- Severe renal disease
- Rare clearing on dialysis
- Toxin or contaminant related to dialysis?
- Coagulation, DVTs, recent failure of a transplanted kidney
- Recent vascular surgical procedure (such as revision of an AV fistula, or angioplasty of a blood vessel) or to have experienced a thrombotic episode (thrombotic loss of a transplant or deep venous thrombosis) approximately two weeks before the onset of the skin changes.
Histopathology

- Thick collagen bundles with surrounding clefts are a prominent finding
- Variable increase in dermal mucin and elastic fibers
- May extend to subcutis and fat
- Florid CD-34 positive dermal dendrocytes
- Factor XIIIa and CD-68 positive mono- and multinucleated cells
Calciphylaxis
Calciphylaxis

- 1-4% of the ESRD population
- Caucasians
- F:M 3:1
- 6 months to 83 years
  - Mean age of 48 years
  - Younger patients with longer duration of renal replacement therapy more predisposed
Increased Risk

- Obesity
  - Thighs, buttocks and lower abdomen
- Glucocorticoid exposure
Pathogenesis

- Chronic renal failure
- Hypercalcemia
- Hyperphosphatemia
- Elevated calcium-phosphate product
- Secondary hyperparathyroidism
- Protein C and protein S deficiencies
Radiologic

- Arborization of vascular calcification within the dermis and subcutaneous tissue
- Common in ESRD—not specific for calciphylaxis
Histopathology

- Incisional biopsy with subQ fat
- Calcification in media of small and medium-sized arterioles
- Intimal hyperplasia and fibrosis
- Mixed inflammatory infiltrate
- Subcutaneous calcium deposits with panniculitis and fat necrosis
- Vascular microthrombi
Differential Diagnosis

- Infectious panniculitis
- Vasculitis
- Thrombotic disorder
Prognosis and Treatment

- Mortality rate 60-80%
  - Leading cause of death is sepsis from infected, necrotic skin lesions
  - Mortality rate is higher in patients with proximal disease than in those with only distal or acral disease

- Supportive
- Total or subtotal parathyroidectomy with autotransplantation
- Avoid glucocorticoids
Overlap NFD and Calciphylaxis

- LE with erythematous, indurated skin
- NFD dermal changes and subQ septa
- Subcutaneous calcification, diffuse and vascular
- Calcification not suspected clinically

J Cutan Pathol 2004;31:247
References