A 42-year-old African American male presented with a four-week history of inflamed nodules with ulcerations on his left arm. Past medical history was significant for renal failure; the patient was currently on dialysis.

Biopsy revealed collection of calcium within the dermis with chronic inflammation. Labs were significant for elevated levels of phosphorus.

Diagnosis: Calciphylaxis

CALCIPHYLAXIS IN REVIEW

A relatively rare condition, calciphylaxis is predominantly seen in chronic kidney failure patients treated with dialysis (uremic calciphylaxis), although the condition has been described in earlier stages of chronic kidney disease and with normal renal function. Incidence of the condition appears to be increasing, although a significant spike in diagnoses in the last decade was associated with the introduction of a specific ICDN code for this condition.

Calciphylaxis is characterized by very painful, ulcerating, and, finally, necrotic skin lesions and is associated with severe pain, non-healing wounds, and recurrent hospitalizations. Lesions (livedo reticularis, reticulate purpura, violaceous plaques, or indurated nodules) that demonstrate poor healing can be complicated by blistering and ulcerations and present the potential for superimposed infections. Treatment itself may be associated with risks for adverse events.

Diagnosis is generally made on the basis of histology, which shows calcifications in the media of small cutaneous arterioles, nerve sheaths, and sometimes, adipose and soft tissues. Deep excision biopsy is preferred for accurate analysis, however, this technique is risky, as it can be associated with ulceration and sepsis.

The one-year mortality in calciphylaxis patients is reported at 45-80 percent; sepsis is the leading cause of death. Higher

Figure 1. Inflamed nodules with ulceration on the left arm of a 42-year-old African American male undergoing dialysis (Top and bottom, left) resolved with treatment (Bottom, right).
rates of mortality are associated with ulcerated lesions. One analysis revealed that mortality rates among calciphylaxis patients were 2.5 to three times higher than average mortality rates for chronic hemodialysis patients. Treatment is aimed at preventing or treating infection and attempting to normalize calcium-phosphate metabolism. Dialysis should be implemented if it is not already being administered. Intravenous sodium thiosulfate has been used. Systemic glucocorticoids may prevent ulceration of early plaques of calciphylaxis.

A 26-year-old obese male presented with a one-week history of palpable hemorrhagic purpura on the extensor surfaces of his legs bilaterally (Figure 2). In addition, he reported joint and abdominal pain. No fever was associated with symptoms. The patient had no significant prior medical problems. Labs were positive for ANA and negative with ANCA, with elevated Hematocrit. Otherwise LFTs, CMP were within normal limits. Urinalysis was positive for red blood cell casts and negative for proteins.

On biopsy, H+E stains showed extensive vascular necrosis with a predominance of neutrophils consistent with small vessel vasculitis/IgA vasculitis (Henoch-Schönlein purpura). Direct Immunofluorescence: Positive granular IgA and slightly weaker IgM, IgG, C5b-9, and fibrinogen deposition in and around the blood vessels. Renal Ultrasound was negative for renal infarcts. The lesions gradually resolved over time with oral prednisone. The patient was also referred to nephrology to monitor renal function.

**Diagnosis:** Henoch-Schönlein vasculitis triggered by dehydration and overexertion

**HENOCH-SCHÖNLEIN PURPURA IN REVIEW**

Henoch-Schönlein purpura is an acute, systemic, immune complex-mediated, leukocytoclastic vasculitis. The vast majority of cases (90 percent) occur in children under age 10. The condition is typically characterized by a triad of palpable purpura (without thrombocytopenia), abdominal pain, and arthritis. Most patients have an antecedent upper respiratory illness. All patients with Henoch-Schönlein purpura develop a purpuric rash, while 75 percent develop arthritis, and 60 to 65 percent develop abdominal pain. Reportedly 40 to 50 percent of those affected by the condition develop renal disease. One recent analysis showed that age greater than 30 years was an independent predictor of renal involvement in Henoch-Schönlein purpura. Risk factors for significant renal involvement increased with every 10-year increase in age. In a significant proportion of children (94 percent) and adults (89 percent), Henoch-Schönlein purpura spontaneously resolves. Supportive treatment with oral prednisone at 1-2mg/kg/day for two weeks has commonly been used to treat associated abdominal and joint symptoms.

**Evan H. Schlam, MD, FAAD, University of Miami; Private Practice, Plantation, FL**

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