A Challenging Case of Leg Ulcers Associated With Retiform Purpura: Cutaneous Oxalosis in the Setting of Primary Type 1 Hyperoxaluria

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Hyperoxaluria, a rare metabolic disorder associated with increased serum oxalate deposits in the skin, kidneys, and other organs, can be primary or secondary.[1] Primary hyperoxaluria (PH) types 1 (PH-1) and 2 (PH-2) are autosomal recessive. The most common type, PH-1, results from deficiency of alanine-glyoxylate aminotransferase causing accumulation of glyoxalate and oxalate.[1,2] The patient in this report had the most prevalent juvenile form of PH-1, characterized by recurrent calcium oxalate kidney stones preceding development of renal failure.[1] In PH-2, a deficiency of D-glycerate dehydrogenase leads to hyperoxaluria and excretion of L-glycerate.[1] These patients typically present with kidney stones, and renal failure is rare.[1] Secondary hyperoxaluria occurs with excessive intake of oxalate in chronic renal disease, and cutaneous lesions are unusual.[1] Skin lesions in PH include retiform purpura, livedo reticularis, acrocyanosis, and peripheral gangrene.[1-3]

A woman in her 30s with rheumatoid arthritis and end-stage renal disease who was undergoing hemodialysis presented for evaluation of nonhealing, painful, reticulated, purpuric, ulcerated plaques and nodules on the thighs of 4 months' duration (Figure A and B). She had recently been diagnosed with calciphylaxis, started sodium thiosulfate with dialysis, and was taking apixaban for deep vein thromboses of the left arm and right leg. Her medical history was notable for juvenile type PH-1. Hyperoxaluria caused her renal failure, and the diagnosis was confirmed by genetic testing. Her serum oxalate level was substantially elevated (37-111 μmol/L).

An excisional biopsy specimen from her thigh revealed ulceration, epidermal necrosis, and pauci-inflammatory, intraluminal occlusion by fibrin in the dermal vessels with hemorrhage (Supplemental Figure 1A, available online at http://www.mayoclinicproceedings.org). Necrotizing lobular fat necrosis with panicular vessel thrombosis but no evidence of intravascular or extravascular calcification was identified (Supplemental Figure 1B). Extensive yellow-brown, oxalate crystals in the dermis and subcutaneous tissue were birefringent under polarized light (Supplemental Figure 2, available online at http://www.mayoclinicproceedings.org). These findings were diagnostic for cutaneous oxalosis rather than calciphylaxis.

The diagnosis of hyperoxaluria can be challenging in the presence of renal failure because similar clinical findings can also be seen in calciphylaxis.[1-3] An excisional or incisional biopsy with adequate subcutaneous tissue is necessary to render the correct diagnosis in these cases. In calciphylaxis, intravascular and extravascular calcification is necessary to confirm the diagnosis because vessel thrombosis within the dermis and subcutaneous tissue can be seen in both calciphylaxis and hyperoxaluria. However, calcium oxalate crystals are seen only in hyperoxaluria.

The patient was not a good candidate for a combined liver-kidney transplant, and her disease was managed with supportive care including wound care, debridement, sodium thiosulfate, hyperbaric oxygen, and apixaban. Early diagnosis is important in hyperoxaluria because the prognosis is poor, and the

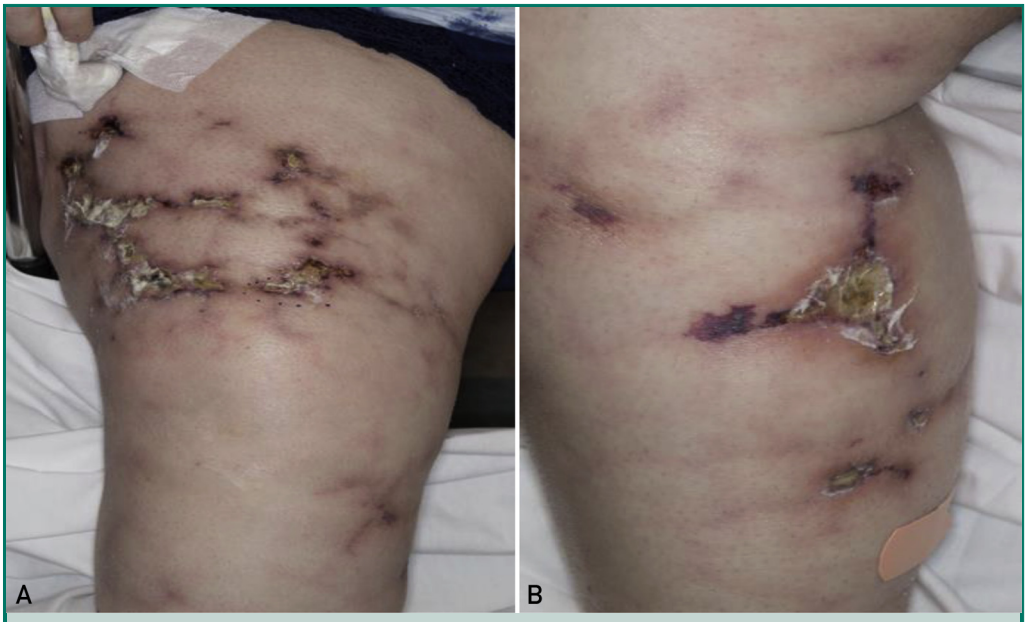

FIGURE. A, Retiform purpura and necrotic ulcers on the thigh and leg. B, Retiform purpura with necrotic ulcers on the leg.

treatment of choice is a combined liver-kidney transplant.[2]

SUPPLEMENTAL ONLINE MATERIAL

Supplemental material can be found online at http://www.mayoclinicproceedings.org. Supplemental material attached to journal articles has not been edited, and the authors take responsibility for the accuracy of all data.

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