



New skin papules

The location of the patient's lesions, as well as her underlying conditions, pointed to the diagnosis.

A 49-YEAR-OLD WOMAN with a history of end-stage renal disease, uncontrolled type 2 diabetes, and congestive heart failure visited the hospital for an acute heart failure exacerbation secondary to missed dialysis appointments. On admission, her provider noted that she had tender, pruritic lesions on the extensor surface of her arms. She said they had appeared 2 to 3 months after she started dialysis. She had attempted to control the pain and pruritus with over-the-counter topical hydrocortisone and oral diphenhydramine but nothing provided relief. She was recommended for follow-up at the hospital for further examination and biopsy of one of her lesions.

At this follow-up visit, the patient noted that the lesions had spread to her left knee. Multiple firm discrete papules and nodules, with central hyperkeratotic plugs, were noted along the extensor surfaces of her forearms, left extensor knee, and around her ankles (FIGURES 1A and 1B). Some of the lesions were tender. Examination of the rest of her skin was normal. A punch biopsy was obtained.

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU TREAT THIS PATIENT?

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FIGURE 1

Papules and nodules on extensor surfaces of the right forearm (A) and left knee (B)



> The acquired form of Kyrle disease is associated with diabetes and renal failure, but there is a lack of data on its pathogenesis.

**Diagnosis:
Kyrle disease**

The patient’s end-stage renal disease and type 2 diabetes—along with findings from the physical examination—led us to suspect Kyrle disease. The punch biopsy, as well as the characteristic keratotic plugs (FIGURE 2) within epidermal invagination that was bordered by hyperkeratotic epidermis, confirmed the diagnosis.

Kyrle disease (also known as *hyperkeratosis follicularis et follicularis in cutem penetrans*) is a rare skin condition. It is 1 of 4 skin conditions that are classified as perforating skin disorders; the other 3 are elastosis perforans serpiginosa, reactive perforating collagenosis, and perforating folliculitis (TABLE^{1,2}).³ Perforating skin disorders share the common characteristic of transepidermal elimination of material from the upper dermis.⁴ These disorders are typically classified based on the nature of the eliminated material and the type of epidermal disruption.⁵

FIGURE 2
Characteristic central hyperkeratotic plugs of Kyrle disease



■ **There are 2 forms of Kyrle disease:** an inherited form often seen in childhood that is not associated with systemic disease and an acquired form that occurs in adulthood, most commonly among women ages 35 to 70 years who have systemic disease.^{3,4,6} The acquired form of Kyrle disease is associated with diabetes and renal failure, but there is a lack of data on its pathogenesis.^{7,8}

■ **Characteristic findings** include discrete pruritic, dry papules and nodules with central keratotic plugs that are occasionally tender. These can manifest over the extensor surface of the extremities, trunk, face, and scalp.^{4,7,9} Lesions most commonly manifest on the extensor surfaces of the lower extremities.

Other conditions that feature pruritic lesions

In addition to the other perforating skin disorders described in the TABLE,^{1,2} the differential for Kyrle disease includes the following:

■ **Prurigo nodularis (PN)** is a skin disorder in which the manifestation of extremely pruritic nodules leads to vigorous scratching and secondary infections. These lesions typically have a grouped and symmetrically distributed appearance. They often appear on extensor surfaces of upper and lower extremities.¹⁰ PN has no known etiology, but like Kyrle disease, is associated with renal failure. Biopsy can help to distinguish PN from Kyrle disease.

■ **Hypertrophic lichen planus** is a pruritic skin disorder characterized by the “6 Ps”: planar, purple, polygonal, pruritic, papules, and plaques. These lesions can mimic the early stages of Kyrle disease.¹¹ However, in the later stages of Kyrle disease, discrete papules with hyperkeratotic plugs develop, whereas large plaques will be seen with lichen planus.

■ **Keratosis pilaris (KP)** is an extremely common, yet benign, disorder in which hair follicles become keratinized.¹² KP can feature rough papules that are often described as “goosebumps” or having a sandpaper-like appearance. These papules often affect the upper arms. KP usually manifests in adolescents or young adults and tends to improve with age.¹² The lesions are typically smaller than those seen in Kyrle disease and are asymptomatic. In addition, KP is not associated with systemic disease.

TABLE

4 perforating skin disorders^{1,2}

Disease	Onset	Distribution	Lesion	Associations
Kyrle disease	Adulthood	Extensor surfaces of extremities (arms and, more often, legs)	Discrete pruritic, dry papules and nodules with central keratotic plugs	End-stage renal disease, type 2 diabetes
Elastosis perforans serpiginosa	Childhood, early adulthood	Neck, flexural areas	Flesh-colored papules with a central plug in annular or serpiginous pattern	Genetic disorders; penicillamine
Reactive perforating collagenosis	Childhood	Hands and arms	Small keratotic papules that enlarge to umbilicated papulonodules, then regress	Genetic
Perforating folliculitis	Early adulthood	Extremities (especially extensor), trunk, buttocks	Small, erythematous follicular papules with central hairs and/or hyperkeratotic cores	Psoriasis

Target symptoms and any underlying conditions

In patients who have an acquired form of the disease, symptoms may improve by treating the underlying condition. For instance, better control of type 2 diabetes may improve symptoms. In patients with end-stage renal disease, a renal transplant can bring complete resolution.¹³

For patients whose Kyrle disease is inherited or whose underlying condition is not easily treated, there are a number of treatment options to consider. First-line treatment includes topical keratolytics (salicylic acid and urea), topical retinoids, and ultraviolet light therapy.^{5,7} Systemic retinoids, topical steroids, cryotherapy, electro-surgery, CO₂ laser surgery, and surgical excision have also been used with some success.^{7,14} Oral histamines and emollients also may help to relieve the pruritus. Lesions often recur upon discontinuation of therapy.

■ Our patient was referred to Dermatology for ultraviolet light therapy. She was also treated with topical 12% ammonium lactate twice daily. Within a few months, she reported improvement of her symptoms. **JFP**

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