

WHAT'S YOUR DIAGNOSIS?

PEER REVIEWED

# What Is This Solitary Ulcerated Nodule?

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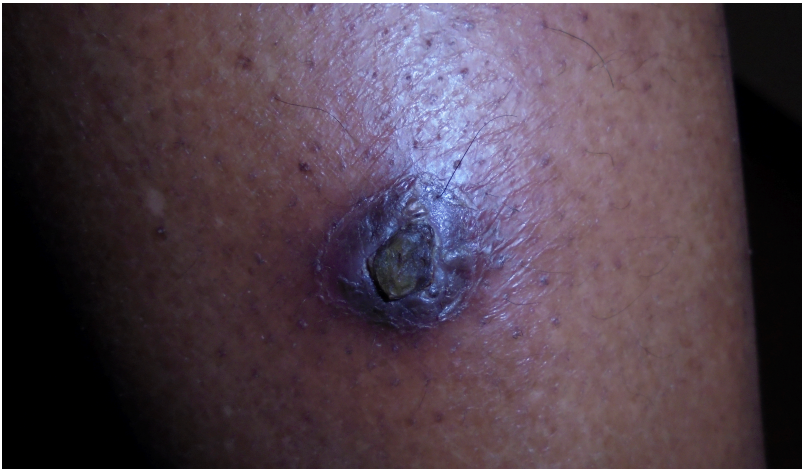
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A 62-year-old African American woman with history of type 2 diabetes mellitus and psoriasis presented with a tender, pruritic, 1.5 × 1.5-cm, ulcerative nodule with peripheral hyperpigmentation and associated hyperkeratotic core on the right lateral lower extremity (**Figure**). The lesion had been present for approximately 2 months, and the patient could not recall any inciting event.



## What's Your Diagnosis?

- A. Prurigo nodularis
- B. Keratoacanthoma
- C. Dermatofibroma
- D. Perforating dermatosis
- E. Arthropod bite

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### Answer: Perforating Dermatosi

More specifically, the patient has a perforating dermatosis known as acquired reactive perforating collagenosis (ARPC).

A shave biopsy was done, the results of which showed thick necroinflammatory scale crust with a focal underlying ulceration and collagen bundles protruding through the ulceration into the crust. Stains for fungi and acid-fast bacilli were negative.

ARPC is an uncommon dermatologic disease process in which altered collagen is extruded through the epidermis.<sup>1-5</sup> It is considered a subtype of the “primary” perforating disorders, which also includes elastosis perforans serpiginosa, perforating folliculitis, and primary (inherited) reactive perforating collagenosis.<sup>1-5</sup>

The diagnosis is based on classic findings of pruritic umbilicated or ulcerated hyperkeratotic papules or nodules, often on the extensor surfaces of the extremities.<sup>1-5</sup> Histopathologic evaluation of these lesions demonstrates transepidermal elimination of degenerated collagen bundles.<sup>1-5</sup> A clinical history of concomitant systemic disease is often elicited, particularly diabetes mellitus (most common) and chronic renal insufficiency, typically as a result of diabetic nephropathy and requiring dialysis.<sup>1-4</sup> Case reports have shown other associated systemic diseases, including hepatic disease, endocrinology disorders (thyroid and parathyroid disorders), hepatocellular carcinoma, Hodgkin lymphoma, acute leukemia, AIDS, tuberculosis, pulmonary aspergillosis, pregnancy, and lupus erythematosus.<sup>1-4</sup>

### Differential Diagnosis

Prurigo nodularis is a relatively common dermatologic disease that occurs secondary to excoriation.<sup>6</sup> The lesions may be distributed on extensor aspects of extremities but can involve the upper back, buttocks, and lumbosacral regions while sparing the mid-upper back (as this area is difficult to reach).<sup>6</sup> They present as intensely pruritic, erythematous or hyperpigmented, firm, dome-shaped papules and nodules with varying degree scale, crust, erosion and/or ulceration.<sup>6</sup> The nodules develop as a result of chronic repetitive scratching or nicking of the

abscesses. The nodules develop as a result of chronic, repetitive scratching or picking of the

skin.<sup>6</sup> Patients typically have pruritus due to dermatologic conditions such as atopic dermatitis, xerosis, and arthropod bites, or may have a psychological condition that leads to repetitive scratching, such as obsessive compulsive disorder, depression, or emotional distress.<sup>6</sup> Histology results demonstrate compact hyperkeratosis with perivascular inflammation and thickened dermal collagen.<sup>6,7</sup>

Keratoacanthoma is a cutaneous malignancy that may represent a variant of squamous cell carcinoma. The lesions occur at any age and are commonly distributed in sun-exposed areas. They typically present as a solitary, dome-shaped, rapidly enlarging papule that evolves over several weeks into a circumscribed, crateriform nodule with a keratotic core. Lesions may resolve slowly months later, leaving an atrophic scar.<sup>6</sup> Histology demonstrates a keratin central core with mild pleomorphism and well-differentiated squamous epithelium.<sup>6,7</sup>

Dermatofibromas are very common benign skin lesions that can occur anywhere on the body but are most often seen on the lower extremities.<sup>6</sup> The lesions are firm, minimally elevated to dome shaped papules and nodules that are often hyperpigmented and dimple when squeezed.<sup>6</sup> The lesions may be idiopathic or associated with trauma or arthropod bites. Histology results demonstrate dermal spindle cell proliferation with trapped collagen.<sup>6,7</sup>

Arthropod bites result in local inflammatory reactions that occur within minutes and usually resolve after several hours; however, reactions may persist for days.<sup>6</sup> The bites often result in erythematous, urticarial papules that can be intensely pruritic and often excoriated from scratching.<sup>6</sup> Lesions may also present as edematous plaques or vesicles and bullae.<sup>6</sup> Especially robust or persistent arthropod bite reactions should trigger evaluation for hematologic malignancy, most commonly chronic lymphocytic leukemia. Histology results demonstrate marked dermal edema with eosinophilic infiltration.<sup>6,7</sup>

The accompanying **Table** summarizes the differential diagnosis and the distinguishing features seen on clinical examination and histopathology tests.

<b>Table. Differential Diagnosis of Perforating Dermatoses</b>	
<b>Condition</b>	<b>Distinguishing Features</b>
ARPC	Umbilicated papules or nodules (typically pruritic) that may eventually form a hyperkeratotic follicular plug, typically in association with a systemic disease (most commonly diabetes and chronic kidney disease). Histology results demonstrate extrusion of altered collagen fibers through an epidermal ulceration.
Prurigo nodularis	Intensely pruritic, erythematous or hyperpigmented, dome-shaped papules and nodules with scale, crust, erosion and/or ulceration. Histology results demonstrate compact hyperkeratosis with

	perivascular inflammation and thickened dermal collagen.
Keratoacanthoma	Hyperkeratotic craters or umbilicated papules and nodules with keratin core/plug that develop rapidly over several weeks and may resolve to become an atrophic scar months later. Histology results demonstrate a keratin central core with mild pleomorphism and well-differentiated squamous epithelium.
Dermatofibroma	Solitary, firm, typically flesh colored (may be hyperpigmented) nodules commonly seen on the lower extremities. Histology results demonstrate dermal spindle cell proliferation with trapped collagen.
Arthropod bite	Erythematous, urticarial papules and nodules that are pruritic and often excoriated from scratching. Histology results demonstrate marked dermal edema with eosinophilic infiltration.

## Treatment

ARPC is often chronic, with associated relapses and remissions. Treatment guidelines for this disease are limited due to a lack of randomized clinical trials. Lesions have a tendency to resolve spontaneously; however, new lesions often may occur. The mainstay of therapy is treatment of the underlying systemic disease.<sup>1-3</sup> Avoidance of skin trauma by controlling pruritus is beneficial in the prevention of new lesions.<sup>5</sup>

Secondary symptoms of pruritus, inflammation, and hyperkeratosis may be treated with topical and systemic modalities. Pruritus and inflammation can be treated with corticosteroids (topical or intralesional) and antihistamines.<sup>1-3</sup> Hyperkeratosis may be treated with topical retinoids and/or urea.<sup>1-3</sup> Some case reports have demonstrated the efficacy of allopurinol, systemic retinoids, methotrexate, and doxycycline in the treatment of refractory cases.<sup>1-3</sup>

Our patient was initially prescribed a topical corticosteroid for pruritus following shave biopsy, but she reported resolution of the pruritus following the procedure.

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