Acquired Perforating Dermatosis: An Innocuous Lesion With Possibly Ominous Implications

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Acquired perforating dermatosis encompasses several specific disease entities occurring in adults that often have overlapping clinical and histologic features. Although chronic renal failure and diabetes mellitus are the most common underlying conditions, several other more rare associations have been noted, including internal malignancy. We describe a case of acquired perforating dermatosis presenting as the first symptom in a 64-year-old man who also was diagnosed to have mild obstructive jaundice due to a periampullary villous adenoma with high-grade dysplasia. In a patient with no other risk factors, the presence of an acquired perforating skin lesion may warrant further investigation to rule out an underlying malignancy.

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Acquired perforating dermatosis encompasses several specific disease entities occurring in adults that often have overlapping clinical and histologic features. Although often forming part of a spectrum, the lesions may be separated into 4 classic perforating disorders: reactive perforating collagenosis (RPC), perforating folliculitis, Kyrle disease, and elastosis perforans serpiginosa. These disorders differ in site and age at presentation, associated or underlying disease states, and type of material extruded through the epidermis. Although chronic renal failure and diabetes mellitus are the most common underlying conditions,[1] several other more rare associations have been noted, including internal malignancy.

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Case Report

A 64-year-old man was admitted for evaluation of jaundice of 4 months' duration. He was referred to the dermatologist with multiple hyperpigmented, hyperkeratotic, mildly pruritic papules over the trunk and extremities of a similar duration. The lesions were not excoriated. Laboratory results on admission included the following levels: serum creatinine, 1.8 mg/dL (reference range, 0.6–1.2 mg/dL); total serum bilirubin, 2.6 mg/dL (reference range, 0.3–1.2 mg/dL); direct serum bilirubin, 1.6 mg/dL (reference range, 0.1–0.3 mg/dL); serum aspartate aminotransferase, 77 U/L (reference range, 10–30 U/L); serum alanine aminotransferase, 92 U/L (reference range, 10–40 U/L); and serum alkaline phosphatase, 471 U/L (reference range, 30–120 U/L). Blood sugars ranged from 124 mg/dL (antecibum reference range, 70–110 mg/dL) to 170 mg/dL (postcibal reference range, 80–140 mg/dL). Hematologic and other serologic parameters, including tumor markers (cancer antigen 19-9) did not reveal any abnormalities. Chest radiograph was unremarkable. Magnetic resonance imaging of the abdomen revealed a 2.0×1.7-cm mass in the periampullary region with dilated pancreatic and common bile ducts and intrahepatic biliary dilatation. Endoscopy of the upper gastrointestinal tract showed bulky ampulla with a superficial 5-mm ulcer, which on biopsy revealed a villous adenoma with high-grade dysplasia.

A skin lesion from the right thigh was biopsied and histopathologic examination showed a crateriform lesion containing parakeratotic and basophilic debris, degenerate connective tissue fibers, and inflammatory cells (Figure 1). Multiple-step sections revealed that the lesion was not folliculocentric and had no hair shaft remnant. The epidermis at the floor was traversed by several vertically oriented fibers that were confirmed to be elastic fibers by special stains (ie, elastic van Gieson, Shikata orcein,

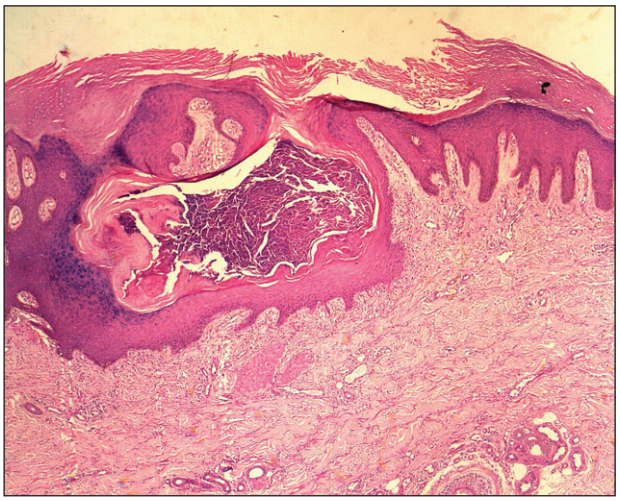

Figure 1. Crateriform epidermal lesion containing parakeratotic and basophilic debris (H&E, original magnification ×25).

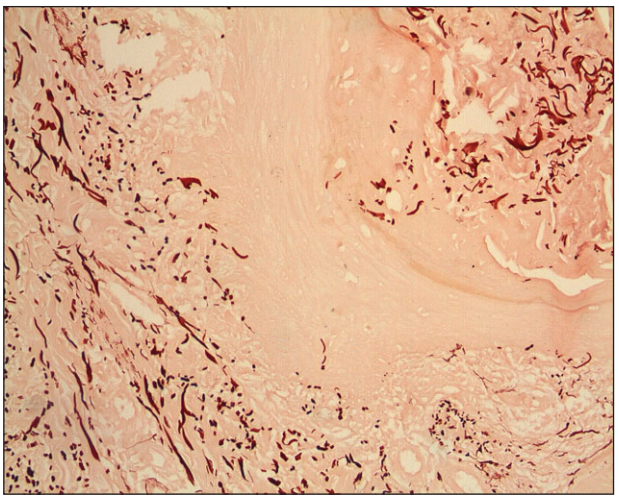

Figure 2. Elastic fibers traversing the epidermis and within the keratotic plug (Shikata orcein, original magnification ×100).

Masson trichrome); the fibers stained positively with elastic van Gieson and Shikata orcein (Figure 2), and negatively with Masson trichrome stain. The absence of collagen fibers precluded a diagnosis of RPC. In Kyrle disease, the keratotic plug does not contain elastic fibers, and the absence of folliculocentricity was against the diagnosis of perforating folliculitis. The clinical findings and histology were not suggestive of elastosis perforans serpiginosa, as there were multiple lesions that were symmetrically distributed, and light microscopy revealed structurally normal elastic fibers. Thus the histologic findings did not fit with any of the classically described perforating dermatoses and a diagnosis of acquired perforating dermatosis was rendered.

Comment

Acquired perforating dermatosis encompasses various types of perforating dermatoses occurring in adults. Clinical and histologic features of the disease are not uniform and may resemble any of the 4 classic perforating disorders: RPC, perforating folliculitis, Kyrle disease, and elastosis perforans serpiginosa.

Reactive perforating collagenosis is a rare disorder with a predisposition to mount an unusual skin reaction to mild trauma with damaged but structurally normal collagen being extruded through the epidermis. The disease may be inherited (presenting in childhood with lesions persisting into adulthood) or acquired. Acquired disease usually presents in adults with underlying diseases as varied as chronic renal failure, diabetes mellitus, herpes zoster, IgA nephropathy, scabies, lymphoma, or rarely underlying carcinoma. The etiology of the disease in these conditions appears uncertain but has been attributed to chronic trauma due to pruritus, a theory that has been refuted by others, as a history of pruritus is not always present.[2] Although RPC classically shows extruded collagen, the presence of elastic fibers also has been reported.[3]

Perforating folliculitis is a folliculocentric disease that shows abnormal keratinization of follicular epithelium, follicular perforation, and exposure of follicular contents to the dermis, which results in frictional damage and transepidermal elimination of connective tissue elements (both elastic and collagen fibers). Histopathologic examination usually reveals distorted curled hair within a dilated follicle with rupture of the follicle at the infundibulum.

Kyrle disease is a controversial entity; it has been recognized as a late-onset genodermatosis[4] or a variant of perforating folliculitis,[5] as perforating lesions in chronic renal failure possess clinical and histologic features that overlap with Kyrle disease and perforating folliculitis. Lesions may be follicular or extrafollicular in location with a predilection for the lower limbs. Histologically, there is a keratotic plug overlying an atrophic invaginated epidermis. Elastic fibers are not present within the plug. When the lesion is folliculocentric, the follicle may rupture at the base.

Elastosis perforans serpiginosa is a rare perforating dermatosis with an arcuate or serpiginous pattern usually confined to one site with transepidermal elimination of abnormal elastic fibers. This condition has been described following penicillamine therapy, with copper depletion in the dermis being postulated as the cause for damage to elastic fibers.

On histology, the elastic fibers are decreased in the dermis, and thick fibers in the papillary dermis are seen penetrating the epidermis. Ultrastructurally, these fibers are convoluted and increased in size.

Although these entities have been described as separate diseases clinically and pathologically, there is much overlap. Cases that cannot easily be subclassified as an acquired perforating lesion of the skin may be called acquired perforating dermatosis. In a review of clinicopathologic features in a series of 22 cases of acquired perforating dermatoses, an underlying systemic disease was present in more than 80% (19/22), most commonly chronic renal failure and diabetes mellitus.[6] Other conditions such as hepatitis, hypothyroidism, tuberculosis lymphadenitis, and a history of a renal transplant also were associated. None of the patients had an underlying malignancy.[6] There have been isolated reports of RPC in patients with internal malignancy[2,7-10] and it has been suggested that the skin lesion may be considered a paraneoplastic condition in these cases. In one case, the skin lesions regressed after the tumor was excised.[7]

In our patient, the jaundice and skin lesions were noticed simultaneously by the patient. Although he presented with only mild jaundice, he reported a history of pruritus, which was likely the inciting event for the skin reaction. Pruritus is one of the most common dermatologic manifestations associated with malignancies,[11] but for unknown reasons, it only rarely results in perforating dermatoses. In a patient with no other risk factors, a perforating dermatosis may warrant further investigation, as this innocuous skin lesion may be the only clue of an underlying malignancy.

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