

JOSEPH F. WALTER, MD, COORDINATOR
La Jolla, Calif

PATHOLOGY QUIZ CASE 1

Edward H. Stolar, MD, the George Washington University Medical Center, Washington, DC

A 57-year-old woman had a four-year history of multiple painful plaques and nodules over the joints of her hands, elbows, and knees. She had occasional joint pain but denied weakness or fatigue. She had been treated with topical fluocinonide 0.05% twice daily for eight months without benefit.

Physical examination revealed hyperkeratotic, hyperpigmented tender nodules as well as a solitary annular plaque with an erythematous elevated border over the joints of her hands (Fig 1). There were

similar nodules over her knees and elbows, and her arms and legs exhibited multiple patches of hyperpigmentation.

Abnormal laboratory findings included an ESR of 56 mm/hr and a hemoglobin level of 10.8 g/dL. An assay for antinuclear antibodies and serum protein electrophoresis findings were normal. Microscopic sections from one of the skin nodules are shown in Figs 2 and 3.

What is your diagnosis?

PATHOLOGY QUIZ CASE 2

Howard K. Steinman, MD, University of California Medical Center, San Diego

A 29-year-old man was seen in the dermatology clinic with numerous, asymptomatic, pigmented penile papules. He had been treated for scabies three months earlier, at which time no penile lesions were noted. There was no history of genital herpes, venereal warts, or arsenic exposure.

Physical examination revealed 15 reddish-brown papules on the penile shaft that ranged from 3 to 7 mm in diameter (Fig 1). Two papules were excised, and representative microscopic sections are shown in Figs 2 and 3.

What is your diagnosis?

PATHOLOGY QUIZ CASE 1



Figure 1.

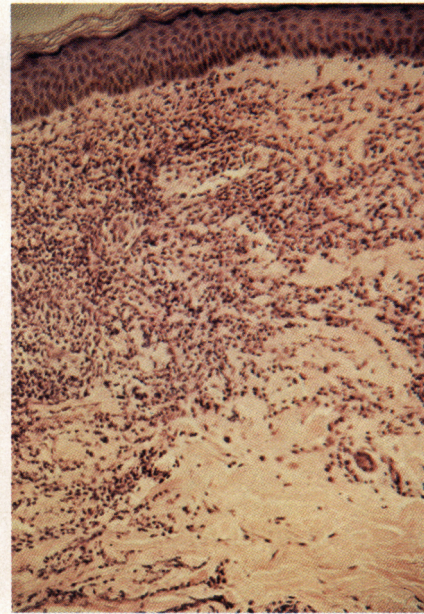


Figure 2.

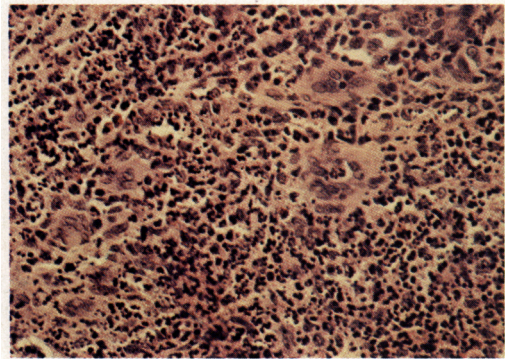


Figure 3.

PATHOLOGY QUIZ CASE 2

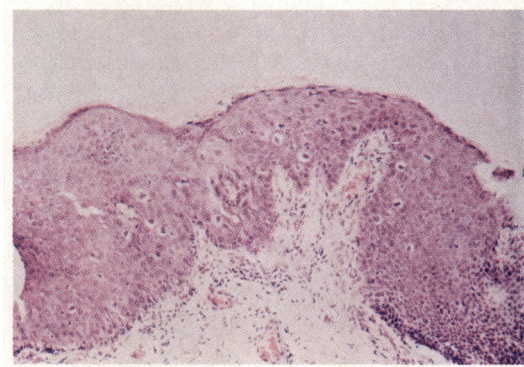


Figure 2.



Figure 1.

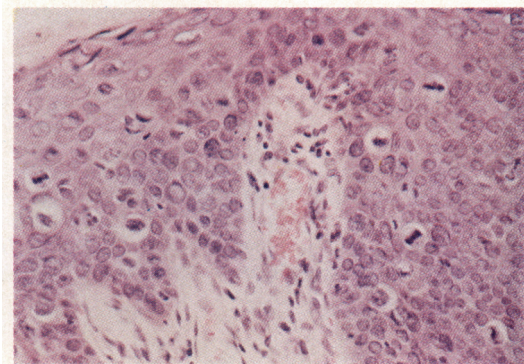


Figure 3.

PATHOLOGY QUIZ CASE 1

Pathology Diagnosis: Erythema elevatum diutinum (EED).

Histologic examination showed the epidermis to be acanthotic, with an underlying grenz zone. The dermis comprised a dense mixed-cell infiltrate of neutrophils, nuclear dust, and eosinophils, with occasional lymphocytes, macrophages and plasma cells. Few blood vessels were identifiable, and their walls showed evidence of fibrinoid degeneration.

Erythema elevatum diutinum is a rare, chronic cutaneous vasculitis. It is characterized by red-purple-brown papules, plaques, or nodules that are distributed symmetrically on the extensor surfaces, particularly over joints. The lesions are often tender. It usually occurs in middle age, but it can occur in childhood as well. The clinical differential diagnosis includes granuloma annulare, Sweet's syndrome, multicentric reticulohistiocytoma, xanthoma, hypertrophic lupus erythematosus, and hypertrophic lichen planus.

The histopathologic findings vary with the stage of the disease. Initially, there is a leukocytoclastic vasculitis of all dermal blood vessels, which is manifested by endothelial swelling, marked prevascular infiltrate of neutrophils and neutrophilic debris, admixed with lymphocytes, histiocytes, eosinophils, and extravasated RBCs. The neutrophilic infiltrate is often found in the interstitium, between collagen bundles. There is also a deposition of eosinophilic material in and around blood vessels. Before this was identified as fibrinoid degeneration, it was referred to as "toxic hyaline."¹ Longstanding lesions may exhibit acanthosis or necrosis of the epidermis. *Extracellular cholesterosis* was a term used to describe the deposition of lipidlike material in areas

of fibrosis within the dermis. The material is doubly refractile and represents cholesterol esters.²

The etiology of EED is unknown. Katz et al³ reported positive reactions to streptokinase-streptodornase skin tests in four of five patients. They also detected HLA-B7 in three of five patients and C1q binding activity, suggesting an immune complex etiology. Fort and Rodman⁴ reported one case in which the condition dramatically improved during pregnancy. Other investigators have demonstrated an association with paraproteinemias.^{3,5}

Treatment with dapsone is justified by the effectiveness of sulfones in neutrophilic-mediated disease, particularly herpetic dermatitis. Dapsone treatment appears to have suppressive but not curative effects, since discontinuation of therapy results in relapse.³ Niacinamide and tetracycline hydrochloride are effective as well.⁶

References

1. Weidman FD, Besancon JH: Erythema elevatum diutinum: Role of streptococci, and relationship to other rheumatic dermatoses. *Arch Dermatol* 1929;20:593-620.
2. Laymon CW: Extracellular cholesterosis. *Arch Dermatol* 1937;35:269-284.
3. Katz SJ, Gallin JI, Hertz KC, et al: Erythema elevatum diutinum: Skin and systemic manifestations, immunologic studies, and successful treatment with dapsone. *Medicine* 1977;56(5):443-455.
4. Fort SL, Rodman OG: Erythema elevatum diutinum: Response to dapsone. *Arch Dermatol* 1977;113:819-822.
5. Kövary PM, Dhonau H, Happle R: Paraproteinemia in erythema elevatum diutinum. *Arch Dermatol Res* 1977;260:153-158.
6. Kohler IK, Lorincz AL: Erythema elevatum diutinum treated with niacinamide and tetracycline. *Arch Dermatol* 1980;116:693-695.

Residents and fellows in dermatology are invited to submit quiz cases to this section. Cases should follow the established pattern and be submitted double spaced and in triplicate. Photomicrographs and illustrations must be clear and submitted as positive color transparencies (35 mm). Do not submit color prints unless accompanied by original transparencies. If photomicrographs are not available, the actual slide from the specimen will be acceptable. Material should be accompanied by the required copyright transfer statement, as noted in "Instructions for Authors." Material for this section should be submitted to Antoinette F. Hood, MD, Department of Dermatology, The Johns Hopkins Medical Institute, 600 N Wolfe St, Baltimore, MD 21205. Reprints are not available.

PATHOLOGY QUIZ CASE 2

Pathology Diagnosis: Bowenoid papulosis (BP).

Microscopic examination showed epidermal hyperkeratosis, focal parakeratosis, hypergranulosis, acanthosis, and papillomatosis. Numerous atypical-appearing keratinocytes, with large hyperchromatic and pleomorphic nuclei, were clustered irregularly throughout the epidermis. Occasional dyskeratotic cells and multinucleate keratinocytes were also seen. The dermis showed a superficial lymphohistiocytic perivascular infiltrate. These histologic features are consistent with a diagnosis of either Bowen's Disease or BP. The sudden appearance of multiple asymptomatic genital papules that clinically seem benign, but histologically resemble squamous cell carcinoma in situ, is characteristic of BP.

Bowenoid papulosis occurs with equal frequency in men and women¹ and almost always begins prior to 40 years of age.^{2,3} It is characterized by small papules that are most commonly located on the penile shaft (and sometimes on the glans) in men and on the vulva, perineum, and perianal skin of women. The lesions are not usually accompanied by symptoms. They may be flesh colored, reddish brown, deeply pigmented, or violaceous, and their surfaces may be smooth, velvety, or verrucous. Patients generally have from two to more than ten lesions, which may be clustered in groups or may coalesce to form plaques.

Clinical features that distinguish BP from Bowen's disease include (1) early onset, (2) multiple lesions, (3) lack of associated symptoms, (4) smaller size and papular nature of the lesions, (5) occurrence in circumcised men, and (6) more common location on the shaft rather than the glans of the penis.^{1,3,4} In no reported case of BP was the diagnosis of Bowen's disease entertained prior to biopsy.

The duration of BP ranges from less than two months to more than ten years. There have been numerous reports of spontaneous regression of lesions,⁵ which is characteristic of the disease.

The cause of BP is unknown, but the following evidence points to a possible viral mechanism: (1) as

many as 32% of the reported cases of BP had a history of genital herpes, 35% had a history of condyloma accuminata³; (2) groups of lesions often appear in the same anatomic area, suggesting an infectious cause; (3) a patient was reported to have concurrent condyloma accuminata and BP, with spontaneous regression of all lesions⁶; (4) histologic features of condyloma accuminata and squamous cell carcinoma in situ have been found in adjacent areas of the same specimen³; and (5) viruslike particles have been detected in BP lesions.⁷

Despite its malignant histologic appearance, the long-term prognosis of BP is good. There has been one reported case of BP progressing to Bowen's disease,⁸ but there have been no reports of BP becoming either invasive or metastatic.

Conservative treatment with close clinical follow-up is indicated for patients with BP. Available treatment methods include surgical excision, electrodesiccation with or without curettage, and cryotherapy. Topical fluorouracil has also been used.^{2,4}

References

1. Arnold HL, Rees RB: Thirty-ninth annual meeting, Meeting Report, American Academy of Dermatology, New York, NY, Dec 6-11, 1980. *J Am Acad Dermatol* 1980;5:484-499.
2. Taylor DR, South DA: Bowenoid papulosis: A review. *Cutis* 1979;27:92-98.
3. Wade TR, Koph AW, Ackerman AB: Bowenoid papulosis of the genitalia. *Arch Dermatol* 1979;115:306-308.
4. Wade TR, Koph AW, Ackerman AB: Bowenoid papulosis of the penis. *Cancer* 1978;42:1980-1983.
5. Berger BW, Hori Y: Multicentric Bowen's disease of the genitalia: Spontaneous regression of lesions. *Arch Dermatol* 1978;114:1698-1699.
6. Steffen C: Concurrence of condylomata accuminata and bowenoid papulosis: Confirmation of the hypothesis that they are related conditions. *Am J Dermatopathol* 1982;4:5-8.
7. Zekickson AS, Praver SE: Bowenoid papulosis of the penis: Demonstration of intranuclear viral-like particles. *Am J Dermatopathol* 1980;2:305-308.
8. DeVillez RL, Stevens CS: Bowenoid papules of the genitalia: A case progressing to Bowen's disease. *J Am Acad Dermatol* 1980;3:145-152.