Diffuse eruption of pigmented papules

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A 33-year-old African American woman presented for evaluation of an itchy eruption of approximately 6 months' duration that began on the hand dorsa before spreading to the arms, ears, and knees. Initial examination revealed clusters of pigmented, 1-mm papules on the hand dorsa,

especially the knuckles, and on the distal forearms. Multiple lesions were present in areas of scratching (Koebner's phenomenon), producing a linear pattern. After the initial evaluation, these lesions continued to spread and involved the forehead (Figure 1), the back of neck (Figure 2), and the distal extremities (Figure 3). On further questioning, the patient stated that she had suffered a flulike illness 2 months previously and that her muscles felt sore, particularly in her arms and legs.

What is your diagnosis?

Figure 1. Clusters of pigmented papules on the forehead.

Figure 2. Multiple pigmented papules on the back of the neck.

Figure 3. Pigmented papules with a linear distribution.

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DIAGNOSIS: Papular mucinosis (scleromyxedema).

PATIENT EVALUATION AND TREATMENT

Relatively few skin conditions produce the classic Koebner's phenomenon; these include psoriasis, warts, and lichen planus/lichen nitidus. In our patient, the clinical appearance of multiple tiny papules in linear patterns on an edematous background is characteristic of the papular form of scleromyxedema. Histopathology of individual papules revealed the presence of mucinous deposits in the mid-dermal region that stained positively with toluidine blue.

The patient's white blood cell count was $9.1 \times 10^3/\mu\text{L}$ with 80% neutrophils, and the platelet count was $469 \times 10^3/\mu\text{L}$. Serum electrophoresis revealed an IgG-lambda monoclonal gammopathy. Further evaluation revealed no lytic bone lesions, with only a mild plasmacytosis in the bone marrow. Urine electrophoresis results were normal.

The patient continued to develop multiple fresh papules, especially on the face, proximal limbs, and upper trunk region. A slowly progressive thickening of the skin was noted, producing an almost sclerodermoid (i.e., orange peel-like) appearance. Skeletal surveys 5 years later again revealed no evidence of metastatic disease. A subsequent serum electrophoresis documented an increase in the IgG spike from 1.3 to 1.8 g/dL. In addition, the patient's globulin level increased to 3.9 g/dL and the albumin-globulin ratio decreased.

Multiple forms of topical therapy were tried, including potent steroids and antipruritics, but all of these were ineffective. The patient was referred to Marvin J. Stone, MD. Due to the progression of the skin disease, he began therapy with melphalan in conjunction with short courses of prednisone. The patient's skin condition improved slightly after the initial course of chemotherapy. A total of 4 courses of melphalan and prednisone were given to her, but minimal improvement was noted with subsequent treatment courses. Cyclophosphamide therapy was contemplated, but unfortunately, the patient was lost to further follow-up.

DISCUSSION

Papular mucinosis/scleromyxedema is a relatively uncommon disease characterized by a diffuse, papular, pigmented eruption due to mucinous deposition in the skin together with an associated paraproteinemia (1–3). This disease is usually chronic and slowly progressive and can involve the gastrointestinal, musculoskeletal, neurological, and cardiovascular systems. The condition was initially described in 1906. It affects men and women equally, with symptoms usually appearing between the ages of 30 and 50 years. Diffuse involvement of the face may produce an almost leonine facies. When the sclerodermoid component predominates, joint movement and ambulation may be difficult.

The disease is invariably associated with a paraproteinemia, usually IgG with lambda light chains. Less frequently, kappa light chains or even a paraprotein of the IgM or IgA class may be found.

Results of treatment are variable. Steroids and melphalan have produced dramatic results in patients with multisystem disease, as have courses of cyclophosphamide, chlorambucil, and interferon alfa (4). In addition, systemic retinoids with or without psoralen plus ultraviolet A therapy may benefit the skin.

The etiology of papular mucinosis/scleromyxedema remains unknown (5). The onset—days to weeks after an acute infection, often streptococcal in origin and producing a low-grade fever/malaise, myalgias, and arthralgias—suggests a possible hypersensitivity reaction. Whether papular mucinosis represents a true plasma cell dyscrasia is unknown, as multiple myeloma rarely, if ever, ensues.

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