## Photoclinic









n 18-year-old girl was referred to our department for evaluation and treatment of a single asymptomatic erythematous plaque she had found on her upper back eight months before. She had been treated unsuccessfully with topical corticosteroids. Physical examination revealed an erythematous plaque composed of waxy confluent papules of 2-3 mm on the upper back. A narrow erythematous ring around the lesion was observed (Figure 1). General physical examination did not reveal any systemic involvement. Medical history revealed

treatment of acne vulgaris by isotretinoin (0.5 mg/kg/day) for two months. In her family history, there was no similar disease.

Results of laboratory tests including blood count, urinalysis, liver and renal function, and thyroid function were within normal limits. Serum protein electrophoresis did not show any evidence for a monoclonal immunopathy. Screening for viral infections was negative. Dermatophytosis was ruled out by direct microscopic examination and fungal culture tests.

A skin biopsy specimen was obtained from the plaque and stained with hematoxylin and eosin (Figure 2) and alcian blue (Figure 3).

What is Your Diagnosis? See the pages 581 – 583 for the diagnosis

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Photoclinic Diagnosis:



Figure 4. Reliability spontaneous regression after two months.

## Discussion

Microscopic section stained with hematoxylin and eosin showed thinning in the epidermis, and fibroblast cell infiltrations were striking in particular, the superficial dermis. Collagen fibers were separated in some places. Infiltrations of lymphocytes and mast cells were also observed in some areas (Figure 2). Alcian blue stained the deposits of mucin in the dermis (Figure 3). Based on the clinical, histologic, and laboratory findings, the disease was diagnosed as plaquelike cutaneous mucinosis (PCM). The patient refused any treatment. During the two-month follow up, relatively spontaneous regression was observed (Figure 4).

Cutaneous mucinoses are a heterogeneous group of disorders in which mucin accumulates in the skin. There is no widely accepted classification for cutaneous mucinoses, but they can be grouped in six types: (1) generalized myxedema; (2) pretibial myxedema; (3) papular mucinosis or localized lichen myxedematous; (4) reticular erythematous mucinosis or plaque-like mucinosis; (5) self-healing juvenile cutaneous mucinosis; and (6) scleredema.<sup>1</sup>

PCM is a rare type of focal cutaneous mucinosis that is characterized by erythematous papules coalescing in a plaque with a reticular pattern on the central chest and upper back. Although PCM and reticular erythematous mucinosis were initially considered to be different, most current authors accept that the two conditions have a single process.<sup>1-4</sup>

Plaque-like cutaneous mucinosis was originally described by Perry et al.<sup>5</sup> in 1960 as a disease composed of confluent papules and was called plaquelike mucinosis. Similar cases with dominating coalescent macules, rather than designated papules. were as reticular erythematous mucinosis by Steigleder et al. in 1974.<sup>6</sup> It has since become apparent that macular and papular lesions can coexist.<sup>2</sup> The clinical presentation is generally asymptomatic, persistent, ervthematous, infiltrated papules, which may either be isolated or coalesce into plaques developing in the midline of the back or chest. It may spread to the upper abdomen. It is also called midline mucinosis.<sup>7</sup> The lesions are occasionally pruritic. Evolution is gradual. The disorder can appear at any ages, but most of the patients are young or middle-aged women.<sup>2,4</sup> In the present case, the plaque composed of erythematous lichenoid papules that started to develop in the interscapular area and then gradually spread.

The exact etiology of PCM is unknown.<sup>8,9</sup> Various theories are discussed such as increased photosensitivity, viral causes, or an immune system disorder.<sup>9–11</sup> Several patients noted suninduced worsening of the condition, lesions have rarely been induced by UVB though.<sup>2,5,12,13</sup> Onset with use of oral contraceptives, menstruations, and pregnancy are other features.

Generally, PCM is not associated with other systemic diseases, but it can appear very similar to mucinosis developing in association with lupus erythematosus. The two share some common features such as flare after exposure to ultraviolet radiation, clinical manifestations, histology, and good response to systemic antimalarials.<sup>14</sup> Coexisting disorders have been reported in association with occasional cases of PCM, perhaps coincidentally.<sup>9, 11</sup>

Histopathologic features of PCM are usually perivascular, and occasionally perifollicular, mononuclear cell infiltration with increased dermal mucin deposition. Direct immunefluorescence is negative. In papular lesions, usually the mucin is fairly conspicuous. Fibroblasts are located in the mucinous deposits.<sup>1,2,15</sup>

PCM can be confused with polymorphic light eruption, drug eruption, lupus erythematosus, Jessner lymphocytic infiltrate of the skin, seborrheic dermatitis, tinea corporis, or superficial psoriasis. For diagnosis, clinical and pathologic correlation is necessary.<sup>1,15</sup>

For the treatment of isolated PCM, systemic or local application of various drugs, such as steroids partly combined with UVB irradiation, systemic antimalarials or cyclosporine, and topical tacrolimus have been reported in the literature.<sup>11,15</sup> Successful treatment of papular mucinosis and lichen myxedematosus by isotretinoin has been reported but isotretinoin has not been used in the treatment of PCM.<sup>16</sup> In the present case, the patient used isotretinoin for the treatment of acne vulgaris for two months just before admission. The lesion was large at that time but partially regressed spontaneously during the two-month follow up. The coincidental use of isotretinoin may have cured the lesion, because isotretinoin is known to treat other mucinosis, papular mucinosis, and lichen myxedematosus. Therefore, it has to be investigated whether isotretinoin may successfully treat patients with PCM.

In conclusion, PCM may appear as a diagnostic challenge, the risk of which is misdiagnosis. This disorder needs to be always kept in mind when there are persistent, erythematous, infiltrated papules or plaques in the midline of the back or chest. Furthermore, isotretinoin may be a potential agent to try for the treatment of PCM.

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