

Papular Mucinosis Associated with Systemic Lupus Erythematosus

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Papulonodular mucinosis (PNM) is a rare variant of lupus erythematosus (LE) eruptions, and PNM is characterized histologically by diffuse dermal mucin without any typical epidermal inflammatory changes. We herein describe a case of papular mucinosis that was characterized by several erythematous papules on the lower back of a 32-year-old man with systemic LE. It is interesting that he didn't display any other skin manifestations of LE such as malar rash, discoid rash and photosensitivity during the previous 2 years. He achieved remission of his PNM without recurrence after 5 months treatment with topical steroids, in addition to receiving systemic antimalarials and steroids.

(*Ann Dermatol (Seoul)* 20(4) 233~236, 2008)

Key Words: Papulonodular mucinosis, Systemic lupus erythematosus

INTRODUCTION

Papulonodular mucinosis (PNM) is a variant of lupus erythematosus (LE), and PNM is characterized by dermal mucin deposition without any of the other histological features of LE. Although this malady was first recognized as early as 1954¹, PNM associated with LE has been reported for fewer than 50 patients. There has been only one reported case of PNM associated with systemic LE in the Korean dermatologic literature². A recent review reported that about 75% of the PNM cases were associated with systemic LE, 20% were associated with discoid LE and 5% were associated with subacute cutaneous LE³. PNM is primarily associated with other cutaneous eruptions in patients with established LE and the PNM fluctuates with the LE disease ac-

tivity⁴. At times, however, PNM can be the only cutaneous manifestation of LE. We herein describe a patient who experienced papular mucinosis as the only cutaneous lesion of his systemic LE.

CASE REPORT

A 32-year-old man presented with a 1-week history of several erythematous papules on the lower back. They had started as tiny skin-colored papules, and these progressively enlarged to form confluent papules. During this time, several new lesions developed. There were no subjective symptoms, and the patient denied any history of injection or trauma. The physical examination showed multiple, relatively well-demarcated, variable sized erythematous papules scattered over his lower back (Fig. 1).

Two years earlier, he had begun to develop arthralgia in his knees and shoulders. He had also noticed joint stiffness in the morning, Raynaud's phenomenon and oral ulcers. He had showed no skin lesions, including malar rash, discoid rash or photosensitivity during the previous 2 years. The significant laboratory values included leukocytopenia ($2,200/\text{mm}^3$), an antinuclear antibody titer of

Received June 8, 2007

Accepted for publication June 1, 2008

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1 : 1280 (a speckled pattern), hypocomplementemia (C3: 53.1 mg/dl [normal range: 88~201 mg/dl], C4: 10.0 mg/dl [normal range: 16~47 mg/dl]) and



Fig. 1. Multiple, relatively well-demarcated, flesh or erythematous colored papules on the lower back.

positivity for antibodies against Ro/SSA, La/SSB and Scl-70. However, the anti-ds DNA antibodies, the hemoglobin level, the erythrocyte sedimentation rate, the kidney and liver function tests and urinalysis were within normal limits. He had been diagnosed with systemic LE and then treated with an antimalarial agent (hydroxychloroquine 100 mg/day) and prednisolone (7.5~10 mg/day) for 2 years, which resulted in substantial improvement.

A 4 mm punch biopsy of skin from a papule on the patient's lower back showed deposition of bluish materials around the sweat glands and between the collagen bundles in the dermis (Fig. 2A, B). The mucinous material was stained with alcian blue at pH 2.5 (Fig. 2C). A scant perivascular lymphocytic infiltrate was observed. The number of fibroblasts in the dermis was not significantly increased. Any histopathologic findings of cutaneous LE were not detected, such as epidermal involvement and vacuolar degeneration at the dermoepidermal junction.

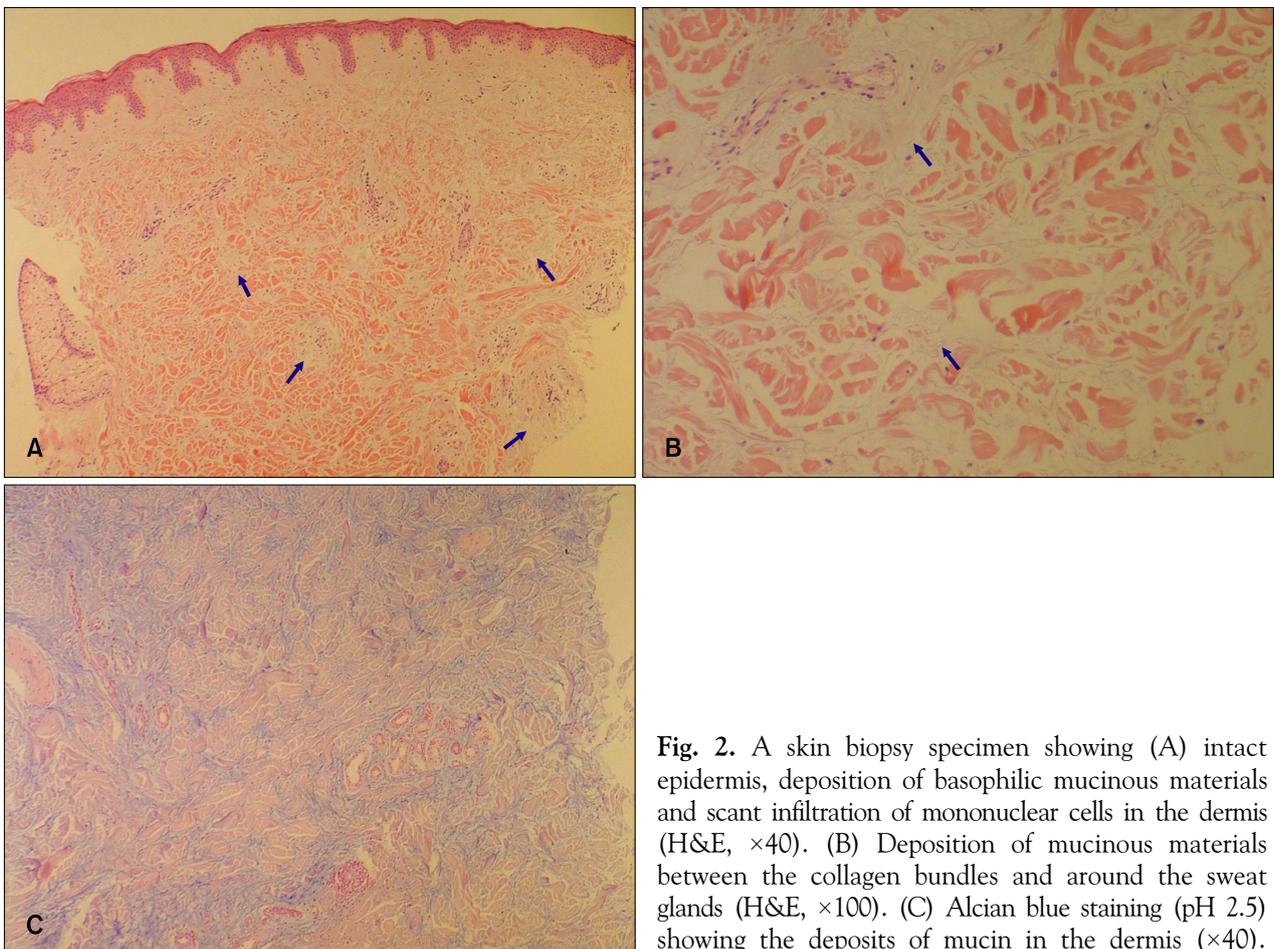


Fig. 2. A skin biopsy specimen showing (A) intact epidermis, deposition of basophilic mucinous materials and scant infiltration of mononuclear cells in the dermis (H&E, $\times 40$). (B) Deposition of mucinous materials between the collagen bundles and around the sweat glands (H&E, $\times 100$). (C) Alcian blue staining (pH 2.5) showing the deposits of mucin in the dermis ($\times 40$).

Based on these findings, the skin lesions were diagnosed as papular mucinosis in a patient with systemic LE. He was treated with topical steroids in addition to receiving systemic antimalarials and prednisolone. After 2 weeks treatment with the medications, he achieved remission of his PNM without recurrence for 5 months.

DISCUSSION

PNM is a distinctive, although unusual cutaneous manifestation of LE and PNM is due to a diffuse deposit of mucin in the dermis. It is clinically characterized by asymptomatic flesh-colored papules and nodules, which occur in the presence or absence of the typical cutaneous lesions of LE. PNM has a predilection for the trunk and upper extremities, but the face and other areas of the body such as the neck and thighs may also be affected². This condition must be distinguished from other cutaneous mucinoses, such as myxedema, reticular erythematous mucinosis, scleredema, alopecia mucinosa and cutaneous focal mucinosis. PNM is histopathologically characterized by mucin deposition between the collagen bundles in the papillary and mid-reticular dermis and there is also mild superficial perivascular lymphocytic infiltrates, whereas the epidermis is normal and it is without basal vacuolopathy. The classic epidermal and dermal inflammatory changes characteristic of LE are not typically seen in PNM.

To the best of our knowledge, of the 33 reported cases of PNM in the English medical literature, 28 were accompanied by other typical cutaneous eruptions of LE, such as discoid lesions, macular erythema, annular atrophic lesions, papulosquamous eruptions, vasculopathic lesions or alopecia^{5,9}. It is interesting that our patient had no skin lesions for 2 years after being diagnosed with systemic LE, and papular mucinosis was the only cutaneous manifestation of his systemic LE. PNM may be a presenting sign of LE³ or it may be the first cutaneous manifestation of LE. Yet for our patient, PNM did not precede the onset of LE, but the PNM occurred after the clinical manifestations of systemic LE.

Several factors may contribute to the pathogenesis of this condition. UV light may aggravate skin lesions. In addition, androgens or other sex-related factors may have a role in the pathogenesis

of PNM because the male-to-female ratio of patients with PNM is much greater than that of patients with systemic LE⁴. Furthermore, serum factors, such as cytokines and circulating antibodies, have been found to upregulate the synthesis of mucin from fibroblasts¹⁰.

Many therapeutic modalities have been proposed to treat this condition, including glucocorticoids, antimalarial agents, retinoids, cyclophosphamide and methotrexate; plasmapheresis and surgical options such as dermabrasion, laser and excision have also been tried, but these are usually unsatisfactory³. However, the skin lesions on our patient responded to medication comprised of topical steroids, systemic antimalarials and prednisolone.

In conclusion, we describe here a patient with papular mucinosis associated with systemic LE, and the patient had no LE-specific skin lesions after being diagnosed with systemic LE.

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