Case Report

Papulonodular mucinosis in a Suspected Systemic Lupus Erythematosus Patient

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We present a 45-years-old suspected systemic lupus erythematosus (SLE) woman who had papulonodular mucinosis (PNM), without other cutaneous LE lesion. The lesions of PNM distributed on both legs which were an unusual location. In addition, the renal involvement was found and suspected from lupus nephritis. The patient was treated with prednisolone, mycofenolate mofetil and chloroquine. After 2 months of follow-up, the renal involvement was improved along with the disappearance of skin lesions.

Keywords: Papulonodular mucinosis, Systemic lupus erythematosus

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A 45-year old Thai female presented with a 9-month-history of multiple painless lumps on both lower legs. She had ischemic heart disease, status post percutaneous coronary intervention for 2 months. Her current medications are amlodipine 5 mg/day, carvedilol 25 mg/day, enalapril 5 mg/day, aspirin 81 mg/day, clopidogrel 75 mg/day, atorvastatin 40 mg/day, ezetimibe 10 mg/day and rabeprazole 20 mg/day.

A physical examination revealed multiple discrete, skin-colored, indurated, well-circumscribed, non-tender, fixed nodules on both shins (Fig. 1). No other active skin lesion was found.

The skin biopsy was done and revealed loosening of collagen fibers in the whole dermis, with light bluish material deposit between collagen fibers (Fig. 2). There was slightly superficial perivascular infiltration with lymphohistiocytes. No epidermal or adnexal involvement was found. Alcian blue stain showed abundant mucin deposits in the whole dermis (Fig. 3). Unfortunately, direct immunofluorescence from the tissue was not done.

Further laboratory investigations revealed anemia and hypoalbuminemia. Blood urea nitrogen, serum creatinine and thyroid function test were normal. Positive anti-nuclear antibody (1:320, homogenous

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Fig. 1 Multiple discrete, skin-colored, indurated, well-circumscribed nodules on both shins.
pattern) and positive anti-Ro-SSA antibody (3+) were found. Anti-La-SSB, anti-ds-DNA and anti-Sm antibodies were negative. Urinary examination revealed proteinuria and microscopic hematuria.

Her clinical pathologic findings were consistent with papulonodular mucinosis (PNM). The diagnosis of suspected systemic lupus erythematosus (SLE) was made, according to the two additional criteria (proteinuria and positive ANA) from the systemic lupus international collaborating clinics (SLICC) criteria 2012[1,2]. Due to the presenting of hematuria, hypalbuminemia and proteinuria, she was suspected to have lupus nephritis[3,4]. The nephrologist decided not to do renal biopsy because the patient had to take aspirin everyday for her coronary problem.

Discussion

Papulonodular mucinosis (PNM) is the primary cutaneous mucinosis mainly occurred in SLE[5-23]. Only one case was reported in progressive systemic sclerosis[24]. PNM could be the presenting sign of LE[5-13] and could reflect the disease activity of SLE[9-11,14]. Almost 80% of LE patients with PNM developed systemic involvement especially the renal and musculoskeletal system[7,12,13,15-18]. In SLE patients, PNM could be the only cutaneous manifestation or occur concomitantly with other cutaneous LE.

Typically, the lesion of PNM is multiple discrete asymptomatic skin-colored or pink to red papules, nodules[5,7,10-12,14,15,17-22] and rarely plaques[6,8,9]. The lesion commonly occurs on upper parts including the back, upper chest, arms, head and neck[5-8,10-17,19,22]. Rare case reports of PNM occurred on lower extremities[6,7]; these patients usually had additional lesions on other parts of the body. In contrast to our patient, she had uncommon PNM distribution solely on the legs without involvement in other areas.

Histopathology shows abundant mucin deposition in the papillary and mid-dermis with normal number of fibroblasts. Slightly perivascular and periadnexal infiltrations with lymphocytes and plasma cells were also seen[6-19,22]. In contrast to tumid LE, this usually has dense perivascular and periadnexal infiltration with abundant mucin deposition[25]. The epidermal change is not seen in PNM as in a typical change in specific LE lesions. Direct immunofluorescence studies may reveal linear or granular deposits of IgG, IgM and C3 at the dermoepidermal junction in some cases[6,8,11,13,15,18,19,22].

Our patient did not fulfill the SLICC criteria for diagnosis of SLE. Nevertheless, she presented with PNM, which is more likely to occur in SLE. In additional to positive ANA and renal involvement, the diagnosis of suspected SLE could give to our patient.

Based on previous reports, the treatments were topical or systemic steroids, anti-malarial drugs and hyaluronidase injection. PNM usually has responses well to systemic steroid either alone or in combination with anti-malarial drugs[7,10,14,16]. Hyaluronidase injection was reported to be effective in large papulonodular mucinosis[23]. The course of disease mainly depends on SLE activity[9,11]. The lesion resolves after SLE has been treated.

The treatments were started with prednisolone 25 mg per day, mycophenolate mofetil 4 g per day, and chloroquine 250 mg per day. After 2 months of treatment, the skin lesions disappeared parallel to the improvement of renal involvement.

Fig. 2 The skin biopsy revealed loosening of collagen fibers in the whole dermis (H&E stain, x200).

Fig. 3 Alcian blue stain showed abundant mucin deposits in the whole dermis (x200).
In conclusion, the present case is the first case report of PNM, uncommonly distributed solely on the legs. The patient had not fulfilled the SLICC criteria for the diagnosis of SLE, but PNM lesions in conjunction with a positive ANA, and renal involvement could diagnose the patient as suspected SLE. The patient responded significantly to the immunosuppressive drugs and chloroquine.

Potential conflicts of interest
None.

References

รายงานผู้ป่วย Papulonodular mucinosis in a suspected systemic lupus erythematosus patient

รู้จักผู้ป่วยมีอาการผื่นแดงกระจายตามผิวหนังต่างๆ ผู้ป่วยมีการวินิจฉัยโรค systemic lupus erythematosus (SLE) ที่ผ่านมา 2 ปี ได้ใช้การรักษาด้วย prednisolone, mycophenolate mofetil และ chloroquine ผลตอบสนองต่อการรักษาด้วย prednisolone, mycophenolate mofetil และ chloroquine