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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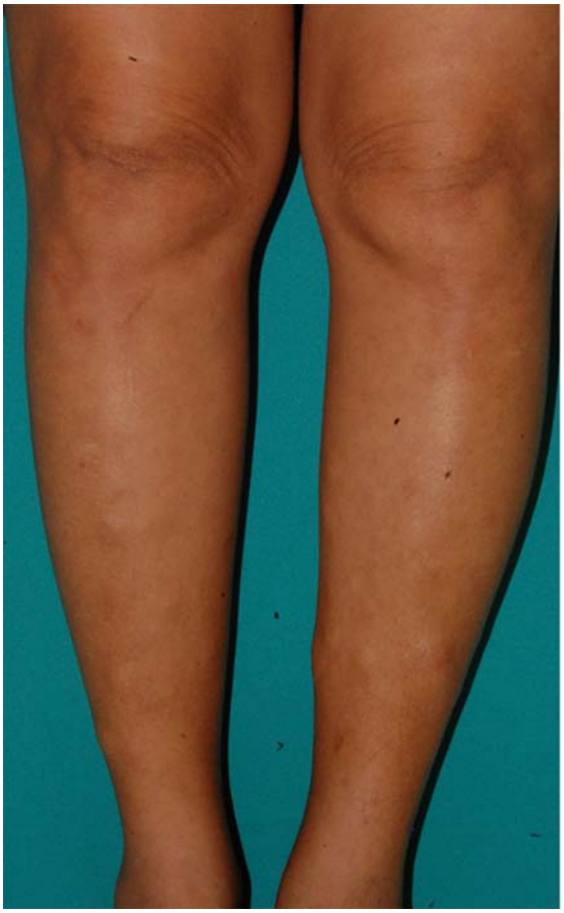

Fig. 1 Multiple discrete, skin-colored, indurated, well-circumscribed nodules on both shins.

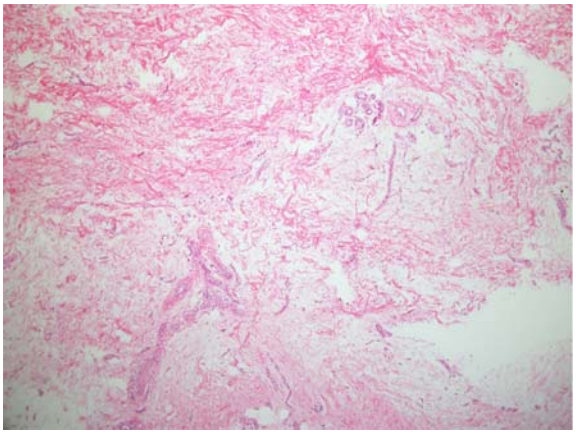

Fig. 2 The skin biopsy revealed loosening of collagen fibers in the whole dermis (H&E stain, x200).

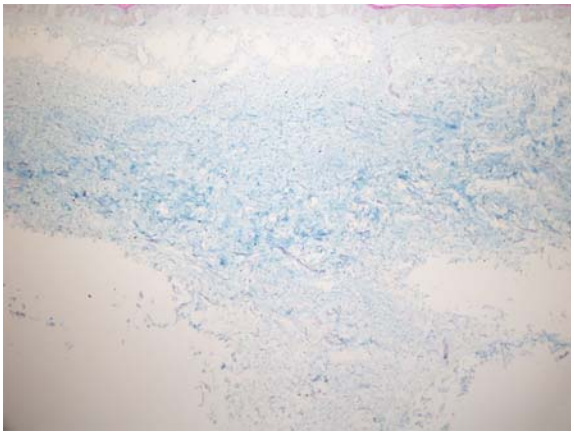

Fig. 3 Alcian blue stain showed abundant mucin deposits in the whole dermis (x200).

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, hypoalbuminemia 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[6,8,11,13,15,18,22].

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.

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

1. Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum 1997; 40: 1725.
2. Petri M, Orbai AM, Alarcon GS, Gordon C, Merrill JT, Fortin PR, et al. Derivation and validation of the Systemic Lupus International Collaborating Clinics classification criteria for systemic lupus erythematosus. Arthritis Rheum 2012; 64: 2677-86.
3. Kodner C. Nephrotic syndrome in adults: diagnosis and management. Am Fam Physician 2009; 80: 1129-34.
4. Weening JJ, D'Agati VD, Schwartz MM, Seshan SV, Alpers CE, Appel GB, et al. The classification of glomerulonephritis in systemic lupus erythematosus revisited. Kidney Int 2004; 65: 521-30.
5. Gold SC. An unusual papular eruption associated with lupus erythematosus. Br J Dermatol 1954; 66: 429-33.
6. Kanda N, Tsuchida T, Watanabe T, Tamaki K. Cutaneous lupus mucinosis: a review of our cases and the possible pathogenesis. J Cutan Pathol 1997; 24: 553-8.
7. Shekari AM, Ghiasi M, Ghasemi E, Kani ZA. Papulonodular mucinosis indicating systemic lupus erythematosus. Clin Exp Dermatol 2009; 34: e558-60.
8. Jackson BA, Norum RA, Lowe L, Fivenson DP. Papulonodular dermal mucinosis in systemic lupus erythematosus. J Rheumatol 1994; 21: 940-1.
9. Pandya AG, Sontheimer RD, Cockerell CJ, Takashima A, Piepkorn M. Papulonodular mucinosis associated with systemic lupus erythematosus: possible mechanisms of increased glycosaminoglycan accumulation. J Am Acad Dermatol 1995; 32: 199-205.
10. Hu S, Wang C, Chan S, Kuo TT. Papulonodularmucinosis associated with systemic lupus erythematosus. Dermatol Sinica 1996: 14; 130-5.
11. Lee WS, Chung J, Ahn SK. Mucinous lupus alopecia associated with papulonodular mucinosis as a new manifestation of lupus erythematosus. Int J Dermatol 1996; 35: 72-3.
12. Bachmeyer C, Chesneau AM, Loirat C, Asensi D, Babinet JM, Blum L. Papulonodular mucinosis in a child with systemic lupus erythematosus. Pediatr Dermatol 2007; 24: 585-6.
13. Lee WS, Chung J, Ahn SK. Plaque and post-auricular nodular mucinosis associated with lupus erythematosus. Int J Dermatol 1997; 36: 367-9.
14. Terao H, Moroi Y, Urabe K, Koga T, Furue M. A case of nodular cutaneous lupus mucinosis. J Dermatol 2003; 30: 341-3.
15. Lowe L, Rapini RP, Golitz LE, Johnson TM. Papulonodular dermal mucinosis in lupus erythematosus. J Am Acad Dermatol 1992; 27: 312-5.
16. Ortiz VG, Krishnan RS, Chen LL, Hsu S. Papulonodular mucinosis in systemic lupus erythematosus. Dermatol Online J 2004; 10: 16.
17. Obermoser G, Sontheimer RD, Zelger B. Overview of common, rare and atypical manifestations of cutaneous lupus erythematosus and histopathological correlates. Lupus 2010; 19: 1050-70.
18. Rongioletti F, Parodi A, Rebora A. Papular and nodular mucinosis as a sign of lupus erythematosus. Dermatologica 1990; 180: 221-3.
19. Hazan C. Papulonodular mucinosis with subacute cutaneous lupus erythematosus. Dermatol Online J 2004; 10: 6.
20. Vazquez GM, Ramirez J, Pena CM, Iglesias GA. A case of lupus associated with papulo-nodular mucinosis. Rev Invest Clin 1994; 46: 63-6.
21. Galofre J, Bielsa I, Casademont J, Herrero C, Cardellach F. Papulonodular mucinosis and late-onset systemic lupus erythematosus associated with Hashimoto's thyroiditis and Sjogren's syndrome. Rev Clin Esp 1989; 184: 446-7.
22. Rongioletti F, Rebora A. The new cutaneous mucinoses: a review with an up-to-date classification of cutaneous mucinoses. J Am Acad Dermatol 1991; 24: 265-70.
23. Maruyama M, Miyauchi S, Hashimoto K. Massive cutaneous mucinosis associated with systemic lupus erythematosus. Br J Dermatol 1997; 137: 450-3.
24. Van Zander J, Shaw JC. Papular and nodular mucinosis as a presenting sign of progressive systemic sclerosis. J Am Acad Dermatol 2002; 46: 304-6.

25. Kuhn A, Sonntag M, Ruzicka T, Lehmann P, Megahed M. Histopathologic findings in lupus erythematosus tumidus: review of 80 patients. J Am Acad Dermatol 2003; 48: 901-8.

รายงานผู้ป่วย Papulonodular mucinosis in a suspected systemic lupus erythematosus patient

รุจิกาญจน์ อัศวรุ่งนิรันดร์, ชุติกา ศรีสุทธิยากร

รายงานผู้ป่วยหญิงอายุ 45 ปี มาโรงพยาบาลด้วย papulonodular mucinosis (PNM) โดยไม่มีอาการอื่น ๆ และนำไปสู่การวินิจฉัยโรค systemic lupus erythematosus (SLE) ที่มีรอยโรคที่ใต้ร่วมด้วยในผู้ป่วยรายนี้พบรอยโรค papulonodular mucinosis ที่ขาทั้ง 2 ข้าง ผู้ป่วยตอบสนองดีต่อการรักษาด้วย prednisolone, mycophenolate mofetil และ chloroquine

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