

# A case report of atypical nodular cutaneous lupus mucinosus

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## Abstract

**Rational:** Nodular cutaneous lupus mucinosus is regarded as a distinctive cutaneous mucinosus deposition with systemic lupus erythematosus (SLE). All typical cases occurred as asymptomatic cutaneous papules, nodules, or plaques on the trunk, upper and lower extremities, and face. Histopathology is mainly revealed abundant mucin deposits among splayed collagen bundles in the dermis. At the same time we can find A the typical clinical manifestations and biological evidence of SLE. Here, we report the first case of nodular cutaneous lupus mucinosus that did not present with any prior symptoms or history of SLE.

**Patient concerns:** We report the first case of nodular cutaneous lupus mucinosus that did not present with any prior symptoms or history of SLE. The patient was 34 years old. One year before admission, nodules began to appear on the elbows, chest, and back, and 2 months before admission erythema occurred on the face. Other notable clinical symptoms were not observed and had no prior history of SLE.

**Diagnoses:** Initially, this patient was misdiagnosed by other clinics as having eczema. After histopathological assessment of skin biopsy and examination of antinuclear antibody signals, the patient was correctly diagnosed with nodular cutaneous lupus mucinosus. Interventions: Followed administration of systemic steroids and hydroxychloroquine.

**Outcomes:** the eruptions quickly disappeared and laboratory indicators improved.

**Lessons:** This case highlights the need for diagnostic vigilance in cases involving papules and nodules initially developing on the chest and elbows in the absence of obvious lupoid symptoms. We recommend a lower threshold for performing histopathological analysis and examination of antinuclear antibody signals in view of the rare but serious possibility of nodular cutaneous lupus mucinosus.

**Abbreviations:** ALT = alanine aminotransferase, AST = aspartate aminotransferase, CRP = C-reactive protein, RNP = ribonucleoprotein, SLE = systemic lupus erythematosus.

**Keywords:** lupus erythematosus, nodular cutaneous lupus mucinosus, pathogenesis

## 1. Introduction

In 1954, Gold first reported that, in patients with systemic lupus erythematosus (SLE), papular and nodular depositions of mucin

Editor: Miao Liu.

All the authors have contributed to this study.

Funding/support: This study was supported by Shandong Provincial Institute of Dermatology and Venereology.

Institutional ethics review board policy is not required for observational case reports that do not alter the patient management. Written informed consent was obtained from patient for the publication.

The authors have no conflicts of interest.

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Medicine (2017) 96:49(e8391)

Received: 13 April 2017 / Received in final form: 1 October 2017 / Accepted: 3 October 2017

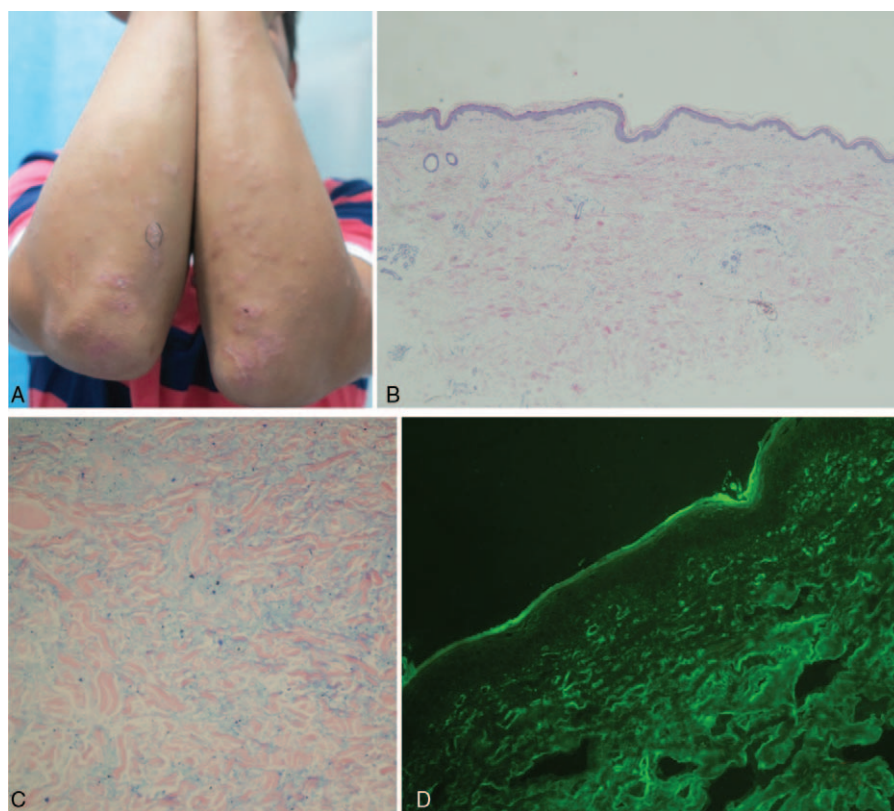
<http://dx.doi.org/10.1097/MD.0000000000008391>

are not limited to sites of eruption.<sup>[1]</sup> Many reports of similar cases followed: in 1985, Nagashima et al<sup>[2]</sup> emphasized the close association of SLE and nodular lesions with mucin deposition, which he named “nodular cutaneous lupus mucinosus.”<sup>[2]</sup> Many similar cases and cases that appear to be variants have also been reported. This is a special case of nodular cutaneous lupus mucinosus, which did not initially manifest as SLE. We believe this case report will ensure an accurate and prompt diagnosis of nodular cutaneous lupus mucinosus.

## 2. Case report

The patient was a 34-year-old male. One year before admission, nodules began to appear on his elbows, chest, and back (Fig. 1A), and 2 months before admission, erythema occurred on his face, without other notable clinical symptoms. Initially, the patient was seen by other clinics who misdiagnosed the case as eczema, prurigo nodularis, and prescribed antihistamine drugs (unknown dosage). However, low efficacy was reported. A few days before admission to our clinic, erythema appeared on both sides of the nose.

Due to first, repeated low efficacy of antihistamine treatment and second, the location of erythema on the cheeks, a region generally afflicted by photosensitive lesions, nodular cutaneous lupus mucinosus was suspected without a clear clinical indication. Examination of a biopsy sample from an eruption on the cheek revealed mucin deposition in the dermis. Laboratory tests were positive for antinuclear antibody (1:000, cytoplasmic pattern),



**Figure 1.** (A) Clinical photographs showing multiple, soft nodules and erythema on the patient's elbows. (B) Histology shows diffuse mucin deposition in the mid and deep reticular dermis. (Hematoxylin and eosin stain; original magnification  $\times 40$ ). (C) Alcian-blue staining and colloidal iron staining showed marked mucin deposition between collagen fibers in the dermis. ( $\times 200$ ). (D) The results of direct immunofluorescence test showed no perivascular deposition of IgG, C3, IgM, IgA.

but negative for anti-double stranded DNA antibody. The serum C3 level was 59 mg/dL (normal range, 90–180 mg/dL), with a C4 level of 5 mg/dL (normal range, 10–40 mg/dL). Other findings included positive anti-RNP (++) , anti-Smith antibody (+), anti-SS-A (+), ribosomal protein (+++), and mitochondrial M2 (+++). Meanwhile, anti-La, anti-Scl-70, and anti-Jo-1 were not detected, which rule out the possibility of Scleroderma. Anti-neutrophil cytoplasmic antibody immunofluorescence, anti-neutrophil cytoplasmic antibody 3, and anti-myeloperoxidase antibodies were not detected. Neither was anti-cardiolipin IgG, or anti- $\beta 2$  glycoprotein IgG, which rules out the possibility of autoimmune diseases other than nodular cutaneous lupus mucinosis. Although lupus anticoagulant testing was negative in this patient, this does not necessary rule out SLE. Complete blood count revealed  $4.61 \times 10^9$ /L white blood cells, 163 g/L hemoglobin, and  $115 \times 10^9$ /L platelets; erythrocyte sedimentation rate was 3 mm/h, and CRP level was 7.09 mg/L. Blood chemistry showed 43 IU/L aspartate aminotransferase (AST), 86 IU/L alanine aminotransferase (ALT; normal range, 0–41 U/L), and 124 alkaline phosphatase U/L ALP124 (normal range, 26–117 U/L). Blood urea nitrogen, creatinine, total protein, albumin, prothrombin time, and activated partial thromboplastin time results were unremarkable. Urinalysis and urine microscopy showed normal results. Thyroid function was normal. Histopathological assessment of a biopsy specimen revealed stark interstitial loosening of collagen bundles in all dermal layers, with moderate perivascular lymphocyte infiltration and no notable alteration in the epidermis (Fig. 1B). Alcian-blue and colloidal iron staining showed marked mucin deposition between collagen fibers in the dermis (Fig. 1C). In

addition, direct immunofluorescence showed no perivascular deposition of IgG, C3, IgM, and IgA (Fig. 1D). On the basis of these clinical and histopathological findings, the patient was diagnosed with atypical nodular cutaneous lupus mucinosis, given the absence of SLE symptoms. Systemic steroids (prednisone 30 mg/day) and immunosuppressants (hydroxychloroquine at 0.2 g twice a day/day) were prescribed. Over the first 2 weeks, all eruptions receded gradually. On the third week, the prednisone dose was reduced to 20 mg/day and then further reduced to 10 mg/day by the fourth week, all without any obvious recurrence of symptoms. From the fifth week, the patient refused to adhere to the treatment and stopped taking prednisone completely. Shortly after discontinuation of treatment, the patient complained of facial erythema, hair loss, and fatigue. He presented to a different clinic out of convenience where he underwent follow-up laboratory testing, which was consistent with our results showing  $6.73 \times 10^9$ /L white blood cells, 173 g/L hemoglobin, and  $112 \times 10^9$ /L platelets. Antinuclear (1:000, speckled pattern) and anti-double-stranded DNA antibodies were positive (242.21 RU/mL; normal range, 0–100) signals. The serum C3 and C4 levels were 0.67 g/L (normal, 0.8–1.8 g/L) and 0.09 g/L (normal, 0.1–0.4 g/L) respectively; other findings were anti-Smith antibody (-), SS-A 29.37 RU/mL (+), SS-B (-), Ribosomal protein P > 200 RU/mL (normal range, 0–20), and mitochondria M2 (+++). Meanwhile, anti-La, anti-Scl-70, and anti-Jo-1 showed no signals. Anti- $\beta 2$  glycoprotein1 was positive (+) and ACL normal. Urinalysis and urine microscopy data were normal. Under the care of the other clinic, prednisone treatment at 30 mg/day for a month resulted in a gradual decrease in most

SLE symptoms such as hair loss. The following month, prednisone dose was decreased to 20 mg/day maintenance treatment for 30 days, then further reduced to 10 mg/day for another 30 days. After 4 months following the initial admission, phone-based follow-up revealed no disease recurrence.

### 3. Discussion

Although before this time, SLE had been represented as having a close association with “nodular cutaneous lupus mucinosis”; many similar cases not limited to this association have been reported.<sup>[3,4]</sup> All previously reported patients had exhibited purely lupoid histological and topical clinical manifestations, including butterfly erythema, fever, arthritis, alopecia, and photosensitivity before the appearance of papules or nodules. However, the patient presented here had papules and nodules on the chest and elbows initially, without any obvious lupoid histological symptoms. Consequently, this patient was misdiagnosed by other clinics as eczema. In our clinic, on the basis of clinical and histopathological findings, as well as positive ANA antibody signals, the patient was correctly diagnosed with nodular cutaneous lupus mucinosis. After administration of systemic steroids and hydroxychloroquine, the eruptions quickly disappeared.

Upon the alleviation of symptoms, the patient refused to adhere to the prescribed treatment. Although the nodules did not reappear after the discontinuation of treatment, other lupus symptoms such as alopecia, facial erythema, and fatigue began to appear. Examination of lupus erythematosus indexes showed ANA and SS-A/ribosomal P protein signals. Blood tests showed that renal and hematologic system remained healthy. A second course of prednisone at 30 mg/day for 1 month resulted in fading of skin lesions and symptoms. Although papulonodular mucinosis (PNM) is an unusual clinical presentation of SLE, the first sign of SLE in this patient indeed presented as cutaneous PNM.

The mechanisms of PNM associated with SLE remain unclear. Interestingly, Pandya et al<sup>[5]</sup> suggest that mucin deposition may be related to increased glycosaminoglycan accumulation by immunoglobulins or cytokines; Kanda et al<sup>[6]</sup> reported that vasculopathy may also be involved in cutaneous lupus mucinosis

development. Zhu et al<sup>[7]</sup> found that collagen and elastin levels within skin lesions differ before and after treatment, which can be used to evaluate treatment efficacy for PNM patients. Genetic mutation may play an important role in autoimmune diseases,<sup>[8]</sup> and provides a compelling framework for future study, as does disease pathogenesis, which our team continues to examine, both to gain a mechanistic understanding and to develop new and effective therapies.

### 4. Conclusion

We report the first case of nodular cutaneous lupus mucinosis that did not present with any prior symptoms or history of SLE for 1 year. The absence of SLE symptoms caused several other clinics to repeatedly misdiagnose this patient with eczema or prurigo nodularis throughout the course of 1 year. Following an expeditious establishment of diagnosis, prompt and regular steroid treatment will most often lead to satisfactory resolution of disease.

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