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LETTER TO THE EDITOR

Numerous Mucin Nodules in a Patient with Seropositive Wide Spread Discoid Lupus Erythematosus

Kozo Yoneda, M.D., Junko Moriue, M.D., Kozo Nakai, M.D., Toshio Demitsu, M.D.[1], Tetsuya Moriue, M.D., Ikumi Yokoi, M.D., Asuka Munehiro, M.D., Hiroaki Dobashi, M.D.[2], Yasuo Kubota, M.D.

Department of Dermatology, Faculty of Medicine, Kagawa University, Kagawa, [1]Jichi Medical University, Omiya Medical Center, Saitama, [2]Division of Rheumatology, Department of Internal Medicine, Faculty of Medicine, Kagawa University, Kagawa, Japan

Dear Editor:

Cutaneous mucinosis comprises of a heterogenous group of pathological disorders characterized by mucin deposition in the dermis. Nodular mucinosis can be an unusual skin manifestation of discoid lupus erythematosus (DLE)[1]. It is associated with increased glycosaminoglycan production by dermal fibroblasts, apparently due to an unidentified serum molecule[2-5]. We describe herein a case of DLE in which cutaneous mucinosis was anticipated because of the sticky gelatinous material from the biopsy sites when the specimens were obtained. Our case had more than 125 mucin nodules on whole body surface area. To the best of our knowledge, this is the first report of a case demonstrating numerous mucin nodules in DLE.

A 38-year-old Japanese man developed cutaneous lesions when he was 25-years-old in 1997. The skin lesions had been treated by his local physician with 0.12% betamethasone valerate ointment. He first presented at our department in June 2006. There was no history of systemic illness. Physical examination showed round, atrophic, erythematous, telangiectatic, and scaly plaques on the face, neck, and all extremities (Fig. 1A). The most dramatic changes were in sun-exposed areas. Irregular encroachment of ear auricular cartilage and overlying skin were noted (Fig. 1A). Blood cell count showed normal values. Serum aspartate transaminase was 50 U/L (normal range 10～35 U/L). Serum alanine transaminase was 110 U/L (normal range 5～40 U/L). Serum gamma-glutamyl transpeptidase was 120 U/L (normal range 0～60 U/L). Serum triglyceride was 190 mg/dl (normal range 30～149 mg/dl). Serum high density lipoprotein was 28 mg/dl (normal range 40～75 mg/dl). Blood urea nitrogen was 8.3 mg/dl (normal range 7～20 mg/dl). Serum creatinine was 0.72 mg/dl (normal range 0.7～1.3 mg/dl). Serum uric acid was 6.0 mg/dl (4.3～8.2 mg/dl). Normal vales for serum C3 and C4 components of complement were found. Antinuclear antibody was positive at 40-fold dilution (normal range is positive at less than 40-fold dilution). Anti dsDNA antibody was 5.0 IU/ml (normal range 0～11.9 IU/ml). Anti-cardiolipin antibodies were negative. The skin lesion was not induced by both ultraviolet A (UVA) and ultraviolet B (UVB) irradiation. There were no signs of serositis, oral ulcers, arthritis, hematological disorder, renal disorder, immunologic disorder, neurologic disorder, or malar rash.

Because we could not find any symptoms other than skin symptoms, DLE was diagnosed and therapy with 0.1% tacrolimus ointment, 0.05% betamethasone butyrate propionate ointment (trunk and extremities), and 0.1% dexamethasone propionate (face) was initiated. Lupus erythematosus (LE) tumidus was excluded because urticaria-like, succulent, erythematous plaques had never been observed. Although the skin lesion was not induced by both UVA and UVB irradiation, it was strongly recommended that the patient avoid sunlight. During the next 3 years, the patient noted that the eruptions waxed and waned.

The patient noticed that skin-colored papules and nodules

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Corresponding author: Kozo Yoneda, M.D., Department of Dermatology, Faculty of Medicine, Kagawa University, 1750-1 Ikenobe, Kitagun Mikicho, 761-0793 Kagawa, Japan. Tel: 81-87-891-2160, Fax: 81-87-891-2163, E-mail: kyoneda@med.kagawa-u.ac.jp

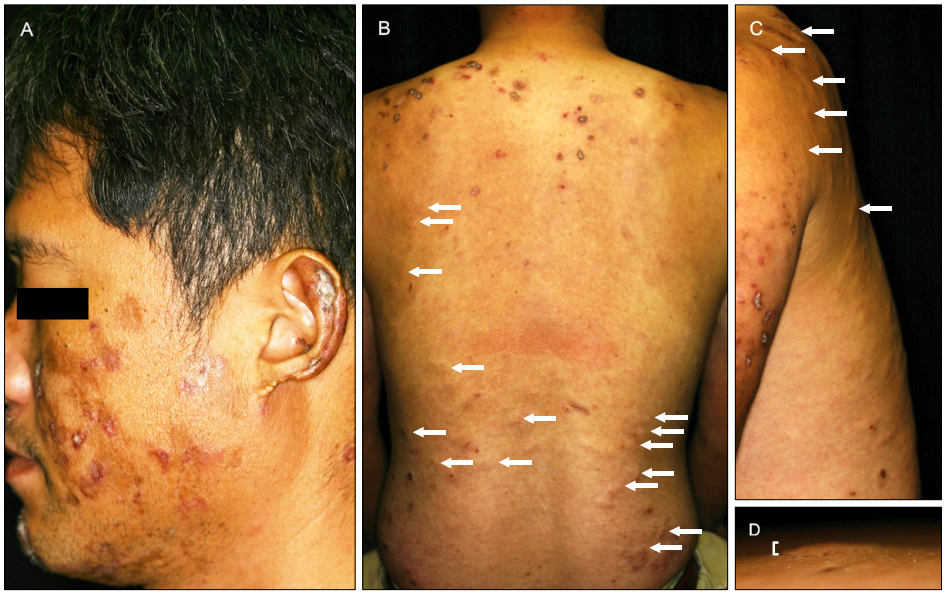

Fig. 1. (A) Round, atrophic, erythematous, telangiectatic, and scaly plaques on the face. Irregular encroachments of the ear auricular cartilage and overlying skin were observed. (B, C) Multiple flesh-colored nodules were observed on the back (arrows). (D) Each nodule was slightly elevated.

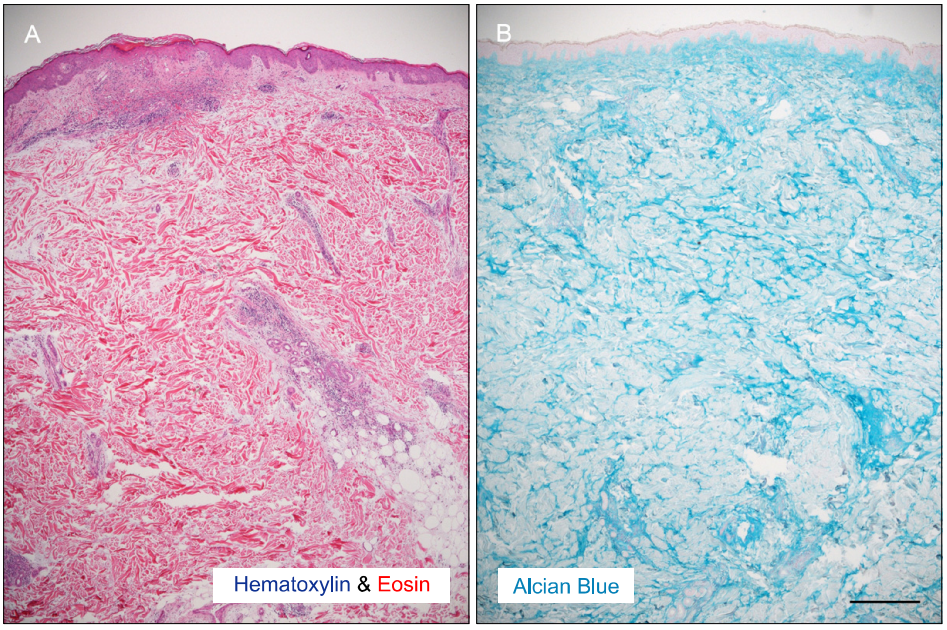

Fig. 2. Histological findings. (A) Focal epidermal atrophy and hydropic degeneration in the basal layer were observed. Papillary edema and a mild perivascular mononuclear cell infiltrate were present in the dermis. The epidermal appendages were normal. Heavy mononuclear cell infiltrate of epidermal appendages was observed (H&E, ×4). (B) Alcian blue staining at pH 4 confirmed the acidic mucopolysaccharide nature of the amorphous material in the dermis. Scale bar: 500 μm (Alcian Blue, ×4).

began to appear on the trunk in September 2008. Examination of the skin in May 2009 showed numerous skin-colored nodules involving mainly in the trunk and extremities (Fig. 1B, C, D). He denied having fever, malaise, and arthralgia. There were numerous nodules on the entire body surface. When a biopsy specimen was taken for light microscopy, the dermis was soft, and mucinous material clung to the biopsy site. The scanning histologic examination of a biopsy specimen from a scaly erythematous lesion on the shoulder showed focal epidermal atrophy and hyperkeratosis. Liquefactive degeneration in the basal cell layer of the epidermis was present. Papillary edema and perivascular mononuclear cell infiltrate were also present in the dermis. The epidermal appendages were normal, but heavy mononuclear cell infiltrate was present around the epidermal appendages (Fig. 2A). Direct immunofluorescence microscopic findings of a specimen from a lesion on the shoulder demonstrated IgA, IgM, and C3 deposition at dermal endothelial cells and band-like IgG deposition along the epidermal basement membrane. Alcian blue staining at pH 4 confirmed the acidic mucopolysaccharide nature of the amorphous material in the dermis (Fig. 2B). At this time point, we still cannot find any symptoms other than skin symptoms. We also could

not observe malar rash, oral ulcers, arthritis, serositis, renal disorder, neurological disorder, hematological disorder, or immunological disorder. We carefully follow up this patient because he may come to systemic lupus erythematosus (SLE) status.

Mucin deposition is a common histopathologic finding in LE but is rarely present in sufficient quantities to produce clinically apparent skin eruptions. To date, 46 cases of papulonodular mucinosis associated with LE were found in the English literature[1-5]. Among them, 34 patients (74%) were diagnosed with SLE, 3 patients (6%) with subacute cutaneous LE, and 9 patients (20%) with DLE. Papulonodular mucinosis differs from other cutaneous eruptions of LE[5]. These lesions present as asymptomatic slightly elevated papules and/or nodules that typically involve the trunk and arms, although the face and other areas of the body may also be affected. Histological examination demonstrates massive mucin deposits in the dermis. Epidermal changes that are typically seen in LE are absent. Several factors are said to contribute to the pathogenesis of papulonodular mucinosis. It has been reported that ultraviolet light exposure can aggravate these lesions[4]. Numerous skin-colored nodules appeared in May in our case. Ultraviolet light exposure may be related to the appearance of many mucin nodules. It has also been suggested that androgen or other sex-related factors may contribute to the pathogenesis[5]. The patient had mild hyperlipidemia. This hyperlipoproteinemia may be related to numerous mucin nodules[6].

In summary, we report a Japanese patient with DLE developed nodular mucinosis 11 years later. Also, he did not have any systemic involvement signs until now.

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