Received: 8 April 2021 | Revised: 18 May 2021 | Accepted: 22 May 2021

DOI: 10.1002/cia2.12190

Journal of Cutaneous Immunology and Allergy

WILEY

CORRESPONDENCE

Anti-MJ/NXP-2 antibody-positive adult-onset dermatomyositis with lichen myxedematosus and endometrial carcinoma

Dear Editor,

Common cutaneous manifestations of dermatomyositis (DM) include Gottron's sign, Gottron's papules, heliotrope rash, poikiloderma, flagellate erythema, and mechanic's hands. Here, we report a case with DM, who was positive for anti-MJ/NXP-2 antibodies, with unique skin eruption as lichen myxedematosus and endometrial carcinoma.

A 53-year-old Japanese woman presented with skin lesions with red papules approximately 3-7 mm diameter on her face, neck, and back (Figure 1A). Gottron's sign was also present (Figure 1B). Histological findings of red papules on her upper back showed vacuolar degeneration of basal cells associated with lymphocyte infiltrates. Lymphocyte infiltrates were seen around the small vessels in the upper dermis. (Figure 1C). Alcian blue staining demonstrated that abundant mucin deposits in the dermis (Figure 1D). Based on the clinical manifestation and histopathological findings, a diagnosis of lichen myxedematosus was made. She did not show calcinosis and generalized subcutaneous edema. She had concurrent weakness of proximal muscles in her upper arms and thighs. The myogenic change was found using electromyogram. Laboratory investigations revealed increased levels of serum creatine kinase, and the presence of anti-MJ/NXP-2 antibodies determined by ELISA.[1] Anti-nuclear, anti-Sm, anti-dsDNA, and anti-RNP antibodies were negative. She was diagnosed as having DM. No signs of interstitial lung disease were obtained. Upon screening of internal malignancies, an endometrial carcinoma was detected. Histopathological findings confirmed the presence of grade 1 endometrioid carcinoma. Abdominal simple hysterectomy, bilateral salpingo-oophorectomy, pelvic lymphadenectomy, omentectomy, and chemotherapy (carboplatin and paclitaxel) were performed. The muscle weakness and skin lesions gradually resolved following the treatment of the malignancy and the oral administration of prednisolone (0.8 mg/kg/day). Prednisolone was tapered over time to be maintained at 0.1 mg/kg/day.

Anti-MJ/NXP-2 antibodies are common in juvenile dermatomyositis.[2] A recent study demonstrated that 1.6% of patients with adult DM or polymyositis (PM) were positive for anti-MJ/NXP-2

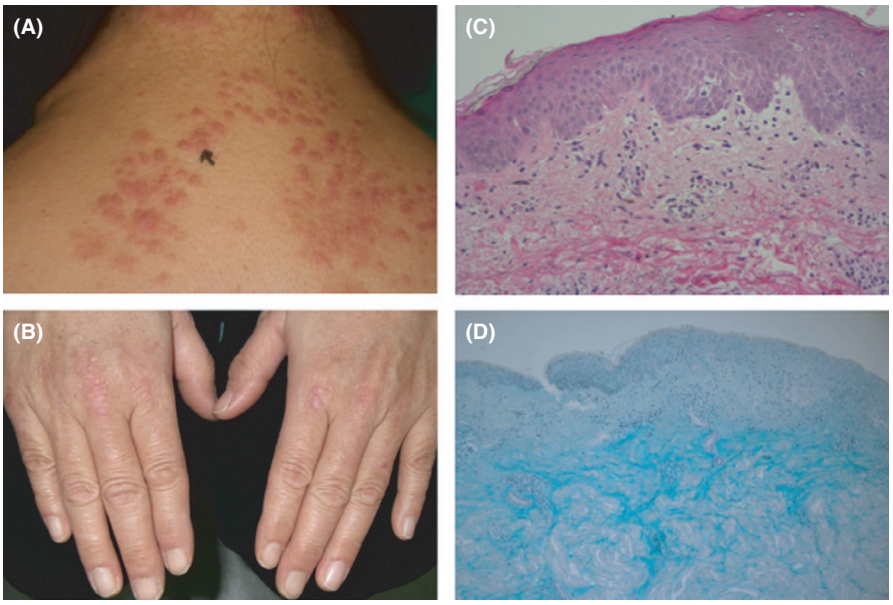

FIGURE 1 Clinical features of our patient. (A) Papules were distributed on her face, neck, and upper back; some of the papules tended to aggregate to form plaques. (B) Gottron's papules were present. (C) Histological findings of a red papule on her upper back showed that vacuolar degeneration of basal cells associated with lymphocyte infiltrates. Lymphocyte infiltrations around the small vessels in the upper dermis were also revealed (Hematoxylin and eosin staining ×200). (D) Mucin deposition was detected in the dermis of skin lesions. (Alucian blue staining ×100)

antibodies in Japan.[3] Moreover, it was shown that DM patients with anti-MJ/NXP-2 antibodies had a significantly higher frequency of calcinosis and generalized subcutaneous edema compared with those without anti-MJ/NXP-2 antibodies.[2,4,5] Our patient showed unique grouping of papules on her face, neck, and back, and she was diagnosed as having DM with lichen myxedematosus. This skin lesion subsided following resection of the comorbid malignancy and treatment with oral prednisolone. Thus, we postulate that localized lichen myxedematosus in this patient occurred in association with DM with anti-MJ/NXP-2 antibodies.

In addition to our case, there were only four case reports in the literature, in which they presented a typical manifestation of lichen myxedematosus with DM.[6–9] Although the pathophysiology of increased mucin deposition remains to be understood, it is postulated that substances circulating in the serum, such as immunoglobulins, autoantibodies, or cytokines, may stimulate glycosaminoglycan synthesis by fibroblasts, leading to the production of mucin and deposition in the skin.[10]

Moreover, this patient had a uterine body neoplasm. It was reported that anti-MJ/NXP-2 antibodies may be associated with adult DM with malignancy.[2,3] This is the first report of the case of anti-MJ/NXP-2 antibody-positive adult-onset DM with lichen myxedematosus and endometrial carcinoma.

DECLARATION SECTION

Approval of the research protocol: N/A.
Informed consent: Informed consent was obtained from the patient.
Registry and registration No. of the study/trial: N/A.
Animal studies: N/A.

CONFLICTS OF INTEREST

The authors declare no conflict of interest. Dr. Shigetoshi Sano is a member of the Journal of Cutaneous Immunology and Allergy Editorial Board. Management of the peer review process, and all editorial decision-making, for this article was undertaken by Editor in Chief.

Kimiko Nakajima MD, PhD[1]
Mayuko Yamamato MD, PhD[1]
Risa Ohsawa MD[1]
Hideki Nakajima MD, PhD[1]
Nagamasa Maeda MD, PhD[2]
Yoshinao Muro MD, PhD[3]
Shigetoshi Sano MD, PhD[1]

[1]*Department of Dermatology, Kochi Medical School, Kochi University, Kochi, Japan*
[2]*Department of Obstetrics and Gynecology, Kochi Medical School, Kochi University, Kochi, Japan*
[3]*Department of Dermatology, Nagoya University Graduate School of Medicine, Nagoya, Japan*

Correspondence
Kimiko Nakajima, MD, PhD, Department of Dermatology, Kochi Medical School, Kochi University, 185-1 Kohasu, Okhocho, Nankoku, Kochi 783-8505, Japan.
Email: nakajimk@kochi-u.ac.jp

ORCID

Kimiko Nakajima https://orcid.org/0000-0002-0341-3252
Hideki Nakajima https://orcid.org/0000-0001-6406-962X
Yoshinao Muro https://orcid.org/0000-0003-2329-8375
Shigetoshi Sano https://orcid.org/0000-0002-9812-0216

REFERENCES

1. Ishikawa A, Muro Y, Sugiura K, Akiyama M. Development of an ELISA for detection of autoantibodies to nuclear matrix protein 2. Rheumatology (Oxford). 2012;51(7):1181–7.
2. Fujimoto M, Watanabe R, Ishitsuka Y, Okiyama N. Recent advances in dermatomyositis-specific autoantibodies. Curr Opin Rheumatol. 2016;28(6):636–44.
3. Ichimura Y, Matsushita T, Hamaguchi Y, Kaji K, Hasegawa M, Tanino Y, et al. Anti-NXP2 autoantibodies in adult patients with idiopathic inflammatory myopathies: possible association with malignancy. Ann Rheum Dis. 2012;71(5):710–3.
4. Albayda J, Pinal-Fernandez I, Huang W, Parks C, Paik J, Casciola-Rosen L, et al. Antinuclear matrix protein 2 autoantibodies and edema, muscle disease, and malignancy risk in dermatomyositis patients. Arthritis Care Res (Hoboken). 2017;69(11):1771–6.
5. Nakagawa Y, Odomari K, Imanaka Y, Katayama I, Ichimura Y, Okiyama N, et al. A case of anti-NXP-2-positive dermatomyositis with generalized subcutaneous edema. J Cutan Immunol Allergy. 2020;3:115–6.
6. Johnson BL, Horowitz IR, Charles CR, Cooper DL. Dermatomyositis and lichen myxedematosus. A clinical, histopathological and electron microscopic study. Dermatologica. 1973;147(2):109–22.
7. del Pozo J, Almagro M, Martínez W, Yebra-Pimentel MT, García-Silva J, Peña-Penabad C, et al. Dermatomyositis and mucinosis. Int J Dermatol. 2001;40(2):120–4.
8. Launay D, Hatron PY, Delaporte E, Hachulla E, Devulder B, Piette F. Scleromyxedema (lichen myxedematosus) associated with dermatomyositis. Br J Dermatol. 2001;144(2):359–62.
9. Wang S. Annular lichen myxedematosus in a patient with dermatomyositis. Int J Dermatol. 2011;50(3):370–2.
10. 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(2 Pt 1):199–205.

How to cite this article: Nakajima K, Yamamato M, Ohsawa R, Nakajima H, Maeda N, Muro Y, et al. Anti-MJ/NXP-2 antibody-positive adult-onset dermatomyositis with lichen myxedematosus and endometrial carcinoma. *J Cutan Immunol Allergy*. 2021;4:173–174. https://doi.org/10.1002/cia2.12190