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LETTER TO THE EDITOR

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A case of anti-NXP-2-positive dermatomyositis with generalized subcutaneous edema

Dear Editor,

Dermatomyositis (DM) is an idiopathic systemic inflammatory disease characterized by proximal muscle weakness and skin manifestations, including Gottron's sign and heliotrope rash. We herein report a case of anti-nuclear matrix protein 2 (NXP-2)-positive DM with generalized subcutaneous edema, which is a rare manifestation of DM.

A 37-year-old Japanese man presented with a 1 month history of erythema on the face involving the nasolabial folds, neck and antecubital fossae, progressive myalgia and proximal muscle weakness, and generalized nonpitting edema (Figure 1A,B,C). His body weight increased by 12 kg within 1 month. He had no medical history. A skin biopsy from an erythematous lesion on the posterior neck showed vacuolar degeneration at the dermoepidermal junction and perivascular lymphocytic infiltration in the upper dermis (Figure 1D). A laboratory examination revealed elevated levels of serum creatinine kinase (13 524 U/L), aldolase (52 U/L), and myoglobin (810 ng/mL). The total protein and the albumin levels were 5.7 and 2.9 g/dL, respectively. The results of urinalysis were normal. The patient was negative for anti-nuclear antibodies (ANA) and myositis-specific autoantibodies against ARS, TIF1-γ, Mi-2, and MDA5, but anti-NXP-2 antibodies were identified by immunoprecipitation-immunoblotting.[1] The electromyographic findings were consistent with inflammatory myopathy, and magnetic resonance

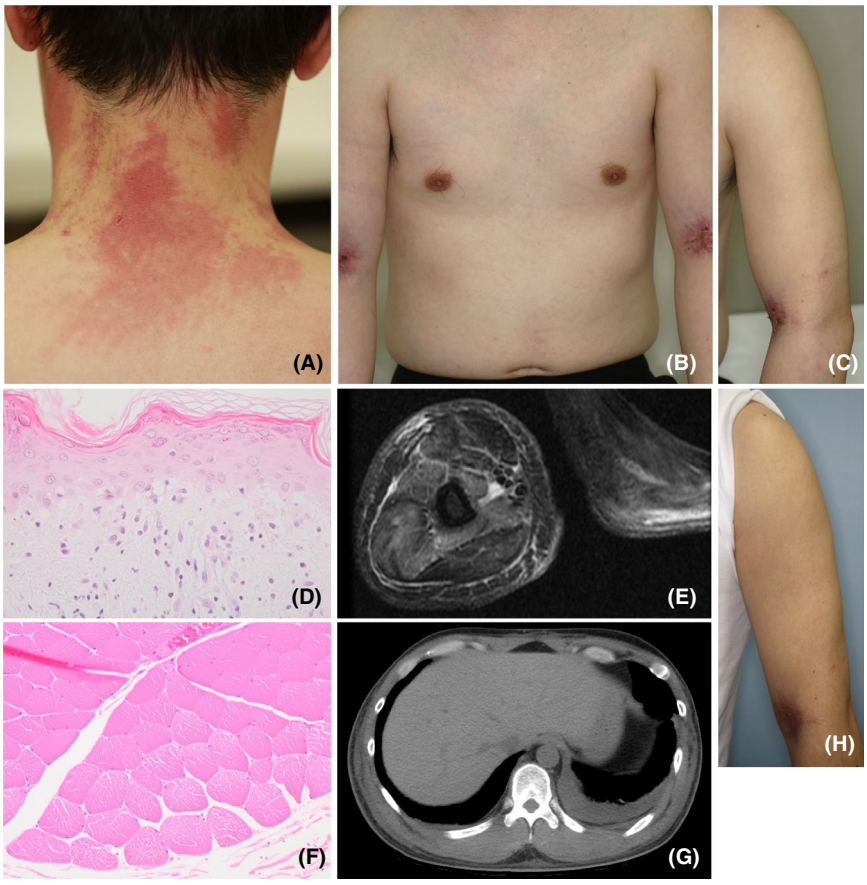

FIGURE 1 A, Erythema on the posterior neck. B, C, Subcutaneous edema on the trunk (B) and extremities (C). D, Hematoxylin-eosin (HE) staining of erythema on the posterior neck showed vacuolar degeneration at the dermoepidermal junction and perivascular lymphocytic infiltration in the upper dermis (original magnification ×400). E, STIR MRI revealed a high signal intensity in the muscles and subcutaneous tissues in the right brachialis. F, A biopsy specimen of the right triceps showed perifascicular muscle fiber atrophy (HE, original magnification ×200). G, Chest CT showed bilateral pleural effusion. H, Subcutaneous edema on the upper extremities disappeared after systemic corticosteroid treatment

imaging (MRI) revealed a high signal intensity in the muscles and subcutaneous tissues in the right brachialis (Figure 1E). A biopsy specimen from the right triceps showed perifascicular muscle fiber atrophy (Figure 1F). Chest computed tomography (CT) showed no abnormalities, with the exception of bilateral pleural effusion (Figure 1G). Internal malignancy was excluded by examinations with abdominal CT and gastroscopy and screening for fecal blood. Based on these findings, the patient was diagnosed with anti-NXP-2-positive DM with generalized subcutaneous edema. No subcutaneous calcinosis was observed. Treatment with oral prednisolone (PSL; 30 mg) significantly improved his muscle weakness, subcutaneous edema, and bilateral pleural effusion (Figure 1H). The abnormalities in the serum levels of muscle enzymes and weight gain returned to normal within 2 weeks, and the PSL dose was gradually tapered without recurrence.

To date, only eight cases of DM with generalized subcutaneous edema have been reported.[2–4] One of these was associated with pleural effusion, similarly to our case.[2] Our patient showed favorable responses to systemic corticosteroid treatment. In contrast, most cases of DM with generalized subcutaneous edema were complicated by dysphagia, were refractory to systemic corticosteroid treatments, and required additional treatments including immunosuppressants and intravenous immunoglobulin.

While no myositis-specific antibodies were reported in the DM cases with generalized subcutaneous edema, an anti-NXP-2 antibody was detected in our case. The clinical features of anti-NXP-2-positive DM include myalgias, dysphagia, calcinosis, malignancy, and peripheral edema,[1,5] although the manifestation of subcutaneous edema in anti-NXP-2-positive DM has not been reported in Japan, which might be because of the different ethnic background.

To our knowledge, this is the first case of anti-NXP-2-positive DM with generalized subcutaneous edema. If generalized subcutaneous edema is observed in DM patients, it is necessary to investigate the possible presence of anti-NXP-2 antibodies.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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