A 1-year-old Japanese boy (Case 1) was referred to our hospital because of muscle weakness and skin ulcers. At the age of 1 year, he lost the ability to walk, although he had started to walk at 11 months. The patient exhibited proximal muscle weakness and muscle grasping pain. The serum creatine kinase level was increased to 136 IU/L (reference range, 41–123 IU/L), erythrocyte sedimentation rate was elevated to 36 mm/h (reference range, 1–10 mm/h), and anti-Jo-1 antibody was positive; magnetic resonance imaging indicated possible myositis of both the femoral muscles. Muscle biopsy revealed histological evidence of a mononuclear inflammatory infiltrate with a perivascular distribution. Furthermore, the patient was noted to have heliotrope rash, Gottron papules, and skin ulcers in the axillary areas (Figure 1). This patient fulfilled two diagnostic criteria for dermatomyositis (Bohan and Peter criteria: definite, and Japan criteria: definite). Accordingly, the patient was diagnosed with juvenile dermatomyositis (JDM).

A 2-year-old Japanese girl (Case 2) was referred to our hospital because of muscle weakness and skin ulcers. At the age of 2 years, she lost the ability to walk, although she had started to walk at 11 months. The patient exhibited proximal muscle weakness, muscle grasping pain, and bilateral knee joint pain. The serum creatine kinase level was increased to 857 IU/L (reference range, 41–123 IU/L), erythrocyte sedimentation rate was elevated to 43 mm/h (reference range, 5–15 mm/h), and anti-Jo-1 antibody was negative; magnetic resonance imaging indicated possible axillary skin ulcers in Case 1 with juvenile dermatomyositis.

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myositis in both the femoral muscles. Muscle biopsy was not performed. Furthermore, the patient was noted to have heliotrope rash, Gottron papules, and skin ulcers in the axillary areas (Figure 2). This patient fulfilled two diagnostic criteria for dermatomyositis (Bohan and Peter criteria: probable, and Japan criteria: definite).1,2 Accordingly, the patient was diagnosed with JDM. 

JDM is a multisystemic disease of uncertain origin that is characterized by chronic inflammation of the striated muscle and skin.3 Skin ulcers occur in 33% and 12% of JDM patients before and after their fifth birthday, respectively.4 Skin ulcers due to vasculitis in patients with JDM are characterized by sharp cut circular lesions, as shown in Figures 1 and 2. The vasculitic skin ulcers in the axilla may become a serious problem in the management of the disease; JDM patients with skin ulcers may have severe and prolonged disease.5,6 Furthermore, skin ulcers should be treated carefully because they can lead to Staphylococcus aureus bacteremia. The axillary skin ulcers in JDM are unique in their clinical appearance. Because these two cases were severe, both patients received prednisolone and methotrexate followed by methylprednisolone pulse therapy for initial therapy. However, to attain remission, Case 1 required cyclosporine owing to resistance to both methotrexate and intravenous cyclophosphamide pulse therapy, while Case 2 required mycophenolate mofetil owing to resistance to methotrexate, intravenous cyclophosphamide pulse therapy, and cyclosporine.

Conflicts of interest

The authors declare no conflicts of interest.

References


Figure 2 Axillary skin ulcers in Case 2 with juvenile dermatomyositis.