A Case of Juvenile Dermatomyositis Manifesting Inflammatory Epidermal Nevus-Like Skin Lesions: Unrecognized Cutaneous Manifestation of Blaschkitis?

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ABSTRACT
Background: Juvenile dermatomyositis is potentially life threatening rare autoimmune illness that mainly affects muscle and skin. Cutaneous features are useful in establishing the diagnosis of this disease.

Case Summary: We report an 8-year-old male juvenile dermatomyositis who presented epidermal nevus-like lesions on the back of the right thigh. Characteristic cutaneous changes such as Gottron’s papules of the hand, heliotrope rash of the eyelids, and poikiloderma-like lesions on the back were observed. Diagnosis of juvenile dermatomyositis was made by positive muscle biopsy and magnetic resonance imaging findings and typical cutaneous manifestations. However, epidermal nevus-like skin lesions, an acquired inflammatory dermatosis that follows Blaschko lines, seen in this case have been rarely reported in the literatures.

Discussion: We would like to report this case and discuss about the significance and pathogenesis of this rare cutaneous manifestation like Blaschkitis in juvenile dermatomyositis.

KEY WORDS
blaschkitis, epidermal nevus-like lesions, Gottron’s papules, heliotrope, juvenile dermatomyositis, poikiloderma

ABBREVIATIONS
JDM, juvenile dermatomyositis; ANA, anti-nuclear antibody; AD, atopic dermatitis; MRI, magnetic resonance imaging.

INTRODUCTION
Dermatomyositis is a multi-system autoimmune disorder primarily affecting the skin and skeletal muscle. Among dermatomyositis, juvenile dermatomyositis (JDM) occurs about 10 cases per year in Japan and manifests characteristic clinical features such as dystrophic calcinosis of the skin, elevated serum IgE or rare association with interstitial pneumonitis or malignancy in addition to typical skin manifestations seen in adult patient with dermatomyositis.1,2 Therefore, it is important to recognize, diagnose and medicate JDM in early stage of the disease. Generally the typical skin rash proceeds before the onset of muscle weakness. However, rare skin manifestations such as inflammatory epidermal nevus-like lesion as documented in the present communication occasionally result in some confusion at the daily practice. Therefore, dermatologist should be careful to recognize the variety of skin manifestations seen in the patients with JDM.

CASE REPORT
The patient is an 8-year old male who had no remark-
able past history and family history. He had had slightly keratinized and itchy skin rash on the anterior side of the lower legs since October 2005, and noticed easy fatigability when he first consulted the dermatological clinic at June 2006. Topical steroid and oral antihistamine treatment failed to improve his complaints. Adversely violaceous papules developed on the each knuckles, extensor surface of elbows, and knees. A liquid nitrogen treatment was enforced them resulting in unfavorable clinical response. Skin rashes extended gradually on the face, extremities and trunk thereafter. Topical vitamin D3 under the diagnosis of pityriasis rubra pilaris, did not show any favorable effect. The patient was consulted to us at October 19, 2006 for further clinical evaluation and additional therapy.

On physical examination, several cutaneous changes were demonstrated as follows. (I) a purplish heliotrope erythema of the eyelids, (II) Gottron’s papules: small erythematous or violaceous, flat papules and small plaques on the dorsa of the knuckles and finger joints (Fig. 1), (III) many scratch marks on the diffuse spreading erythema on the chest and abdomen, (IV) poikiloderma-like lesions with distinct border and scratch marks on the back (Fig. 2), and (V) erythema with white silver scale on the extensor surface of elbows, and knees, keratinized and violaceous papules with zosteriform-distribution on the back of the right thigh mimicking...
Epidermal nevus (Fig. 3). Hematological examination showed following abnormal data: WBC $4.28 \times 10^3/\mu l$ (eosinophil 16.3%), aldolase 9 U/l, anti-nuclear antibody (ANA) ×160 (ANA pattern HO+SP), IgE-RIST 2500 IU/ml, IgE-RAST (house dust 100<). Serum anti-Jo-1 antibody was negative. Histological examination of skin biopsy specimen from the right hand showed hyperkeratosis without parakeratosis, liquefaction degeneration and lymphocytic infiltration surrounding vessels (Fig. 4). Alcian blue-stain showed deposition of mucin in dermis (data not shown). Unfortunately, because of an absence of parental consent, a skin biopsy of the inflammatory epidermal nevus-like skin lesion was not performed.

In this case, the patient had chronic eczema, peripheral blood eosinophilia and high serum IgE level. Based on these findings, differential diagnosis of atopic dermatitis (AD) might be considered. However skin rash are not typical for AD and histological findings are consistent with dermatomyositis rather than those of AD. Finally, we diagnosed him as JDM. Muscle biopsy specimen from the right biceps brachii muscle showed infiltration by inflammatory cells surrounding blood vessels and atrophy of muscle fiber (Fig. 5). A T2-weighted magnetic resonance imaging (MRI) scan showed high-intensity lesion symmetrically in each quadriceps femoris muscles. Gower’s sign, dysphagia and dysarthria were not observed, but tonus of proximal muscles was diminished. A chest roentgenogram did not suggest interstitial pneumonitis signs.

We have administrated oral prednisolone 0.5 mg/kg/day and methotrexate 15 mg/m²/week and used topical steroid for skin rash since November 15, 2006. Currently, skin rash including verruciformis papules on the posterior site of the right thigh almost disap-

**Fig. 4** Skin biopsy specimen from the erythema on the right hand (H-E stain, magnification: ×40). Histological examination of skin biopsy specimen from the right hand showed hyperkeratosis without parakeratosis, liquefaction degeneration and lymphocytic infiltration surrounding vessels.

**Fig. 5** Muscle biopsy specimen from the right biceps brachii (H-E stain, Scale Bar: 0.2 mm). Muscle biopsy specimen from the right biceps brachii muscle showed infiltration by inflammatory cells surrounding blood vessels and atrophy of muscle fiber.

peared. Weakness of the muscle strength also improved, and serum aldolase level returned normal level.

**DISCUSSION**

In this case, the violaceous rash on the posterior of right thigh was clearly different from other typical rash seen in JDM. The rash seems to distribute along Blaschko line and looked like epidermal nevus or blaschkitis. It was noteworthy that this skin lesion began to appear with other characteristic rash of JDM and clearly disappeared after systemic therapy for JDM. From this characteristic clinical course, we consider that the epidermal nevus-like lesion seen in this case is a variant of flagellate erythema. There are some reports of the flagellate erythema mainly as a side effect of bleomycin. A few cases appeared it in patients with DM without bleomycin therapy. The manifestation of flagellate erythema was described as linear violaceous streak, pruriginous lesion and zebra-like stripe eruption. However, a skin biopsy of the lesion was not taken because the patient and his mother disagree to take the skin biopsy, we can never deny the diagnosis of epidermal nevus. Histopathology of flagellate erythema in DM is non-specific. Physical injury, minor trauma and sun exposure have been speculated as causative agents of flagellate erythema. In this case, flagellate erythema might be induced by scratching. Interestingly, we found a report that DM patients with flagellate erythema were positive for anti-155/140 kDa nuclear proteins antibody more frequently than DM patients without flagellate erythema. Internal malignancy was found more frequently in anti-155/140-positive DM patients than anti-155/140-negative DM patients while prevalence of interstitial pneumonia in anti-155/140-positive DM patients was lower than anti-155/140-negative DM patients. However, the antibody
prevalence in JDM patients under 13 years old has not been reported. Because these complications might determine the prognosis of DM, it seems to be favorable that anti-155/140 should be measured if the JDM patients have flagellate erythema. It is advocated that flagellate erythema-associated DM may be more frequent than reported in the literature. Identically, flagellate erythema-associated JDM may be more frequent than reported. The analysis of the relationship between flagellate erythema and anti-155/140 is very interesting and additional study is expected in the future.

ACKNOWLEDGEMENTS

The Japanese version of this work, which did not undergo peer review, was published as the report in the Practical Dermatology. The article title of this English version has been modified because this is more informative for the readers. This English version does not alter the data or interpretations of the Japanese version.

REFERENCES