Superimposed Segmental Manifestation of Juvenile Amyopathic Dermatomyositis in a 9-year-old Boy

Isil Bulur, Hilal Kaya Erdogan, Zeynep Nurhan Saracoğlu, Rudolf Happle1, Funda Canaz2

Abstract
Dermatomyositis is a well-known autoimmune disorder. On the other hand, juvenile amyopathic dermatomyositis (JADM) occurs rather rarely. Here, we report an unusual case in a 9-year-old Turkish boy showing a unilateral linear inflammatory skin lesion that was followed, after 16 months, by the appearance of bilateral disseminated features JADM.

Keywords: Children, juvenile dermatomyositis, superimposed segmental dermatomyositis

Introduction
Superimposed segmental dermatomyositis was defined as an initial severe involvement arranged in a segmental pattern, with subsequent appearance of disseminated lesion.1 We present a case of superimposed linear manifestation of juvenile amyopathic dermatomyositis (JADM) and emphasize that this linear presentation is usually observed as an initial finding in juvenile dermatomyositis (JDM).

Case Report
A 9-year-old boy was presented to our clinic with red-white discoloration in a Blaschko-linear distribution on the left lateral forehead accompanied by mild itching which started 10 months ago. There was nothing of significance in the patient’s personal and family history. The dermatological examination revealed erythematous plaque with an occasional hypopigmented appearance similar to the linear morphea that extended from the left frontal region to the scalp [Figure 1]. The histopathology of the skin biopsy from the lesion revealed interface changes in the epidermis and perivascular, perifollicular, perieccrine lymphohistiocytic infiltration in the dermis together with interstitial mucin accumulation [Figure 2]. The patient was evaluated for collagen tissue disease with the result of the biopsy, but no additional systemic or cutaneous finding was found except for the rash on the face. Full blood and biochemistry examinations were within normal limits. The sedimentation rate was 21 mm/h, C-reactive protein was 0.32 mg/dl, antinuclear antibody was positive, and anti-dsDNA was negative, while anti-Jo-1, anti-Sm, anti-SCL-70, anti-SS-A, anti-SS-B, and anti-RNP were all within normal limits. Topical steroid treatment and alternating topical pimecrolimus treatment were started as a result of this finding. The patient was admitted to our clinic with new skin lesions 16 months later after his first lesion. Dermatological examination revealed malar erythema, erythematous scaly plaques on bilateral knees, elbows and the dorsal part of the fingers, and periungual telangiectasia [Figure 3]. Although the patient had increased symptoms of diffuse muscle pain...
and systemic steroid treatment together with a topical steroid.

**Discussion**

JADM is a rare variant of JDM characterized by only the classical cutaneous findings of dermatomyositis without the clinical and laboratory findings of muscle involvement for at least 6 months. According to the data in the literature, cutaneous findings are preliminary in initial symptoms of JDM patients. Recently, JDM cases presented with initial Blaschko-linear involvement have been reported in the literature. These cases in the literature have been defined as superimposed segmental dermatomyositis. In our patient, histopathological examination of the initial linear skin lesion was consistent with dermatomyositis. Sixteen months later, he developed malar erythema, Gottron papules, psoriasiform rash on the elbows and knees, and periungual telangiectasia which is why the case can be taken as a further example of superimposed segmental dermatomyositis as documented in several previous reports.

**Conclusion**

It is possible to miss the diagnosis of the rare disorder of JADM with this unusual initial presentation. We think that superimposed segmental dermatomyositis is more common than previously reported. In particular, it should be borne in mind by pediatricians and pediatric dermatologists that superimposed segmental dermatomyositis is a juvenile disorder.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

**What is new?**

Blaschko-linear cutaneous involvement is the initial lesion of superimposed segmental dermatomyositis.

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