# Periorbital rash and scaly plaques in a 13-year–old boy

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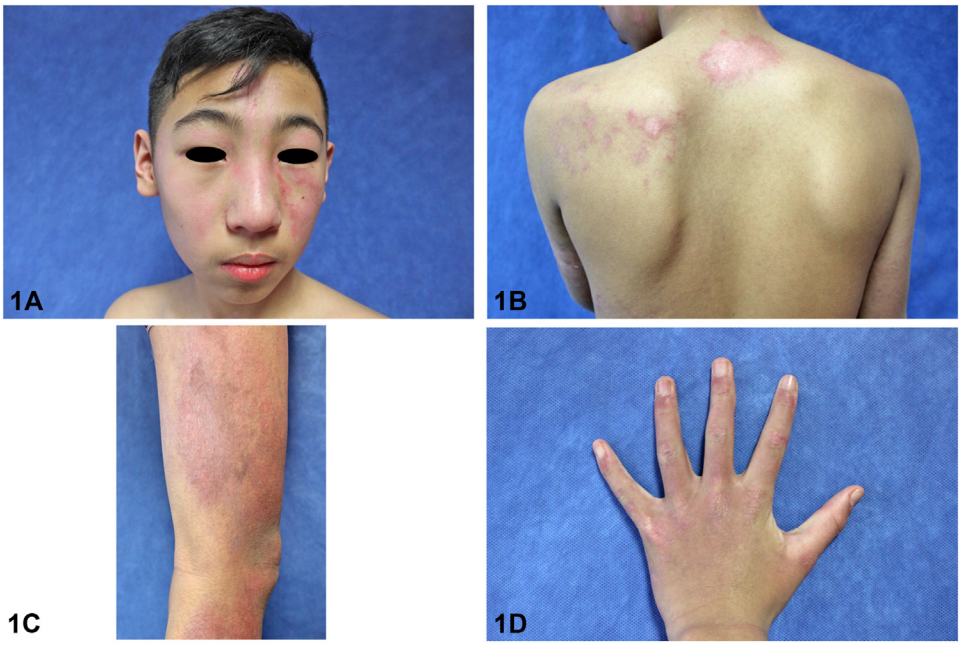


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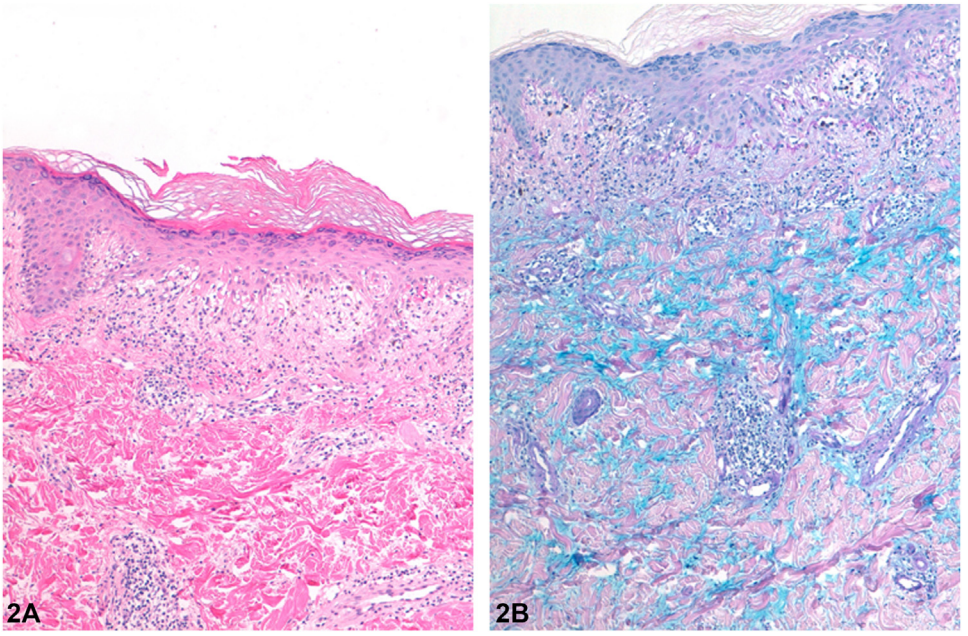


## INTRODUCTION

A 13-year–old boy presented with a 6-month history of a pruritic, periorbital, reddish-purple rash (Fig 1, *A*) and pink-violaceous, noninfiltrated papules and plaques, some with associated scale, on his upper back, extensor arms, and lateral thighs (Fig 1, *B* and *C*). He had similar violaceous lesions with lichenoid quality on the interphalangeal joints of both hands, cuticular overgrowth, and nailfold telangiectasia (Fig 1, *D*). No complaints of weakness, malaise, loss of energy, fever, dysphagia, or respiratory symptoms were reported. Proximal and distal muscle strength were normal.

A skin biopsy specimen revealed vacuolar interface dermatitis with superficial perivascular lymphocytic infiltrate, dermal mucin deposition, and edema (Fig 2, *A* and *B*). Antinuclear antibodies were positive at a titer of 1:160. Laboratory data, including levels of creatine kinase, aldolase, alanine transaminase, aspartate aminotransferase, lactate dehydrogenase, and acute phase reactants, were in the normal ranges. Magnetic resonance imaging showed no evidence of muscle inflammation.

**Question 1: What is the most likely diagnosis?**

**A.** Juvenile-onset systemic lupus erythematosus (SLE)

**B.** Atopic dermatitis

**C.** Allergic contact dermatitis

**D.** Clinically amyopathic juvenile dermatomyositis (CAJDM)

**E.** Systemic sclerosis (SSc)

**Answers:**

**A.** Juvenile-onset SLE — Incorrect. Following the 2019 (American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria, a diagnosis of SLE cannot be made without systemic involvement and with normal laboratory data. Moreover, in SLE, facial lesions would present as a red, butterfly-shaped rash over the cheeks and nose and between the interphalangeal joints.

**B.** Atopic dermatitis — Incorrect. The absence of muscular involvement leads to a differential diagnosis with papulosquamous dermatoses, such as atopic dermatitis. However, the clinical distribution of cutaneous lesions and the absence of spongiotic dermatitis in the skin biopsy specimen are against the diagnosis of atopic dermatitis.

**C.** Allergic contact dermatitis — Incorrect. Eyelid skin is susceptible to the actions of irritating or allergenic agents; therefore, it can frequently be affected in the context of contact dermatitis. However, violaceous color and cutaneous lesions on both hands cannot be related to an exogenous

agent. A cutaneous biopsy would show spongiotic dermatitis.

**D.** CAJDM — Correct. CAJDM is defined by the presence of hallmark skin lesions of dermatomyositis (heliotrope rash, Gottron papules, nailfold capillary changes, and poikiloderma involving the "V" of the chest and the upper back), with no clinical evidence of muscle disease.[1] It represents less than 1% of all juvenile idiopathic inflammatory myopathies (JIIMs).[2] Cases of CAJDM can be classified as amyopathic (no markers of myositis) or hypomyopathic (laboratory, electrophysiologic, or radiologic evidence of myositis).[3] Based on the 2017 (American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for idiopathic inflammatory myopathies, the diagnosis could be assessed without a muscle biopsy.

**E.** SSc — Incorrect. Cutaneous involvement in SSc includes skin thickening, digital ulcers, megacapillaries in the proximal nailfold, calcinosis, and Raynaud phenomenon. However, papulosquamous plaques in the context of this connective tissue disorder are not usually found.

### Question 2: What are the most frequent myositis-specific antibodies in patients with CAJDM?

**A.** Anti-3-hydroxy-3-methylglutaryl coenzyme A reductase (HMGCR)

**B.** Anti-p155/140 (anti-transcription intermediary factor 1-gamma [TIF]$\gamma$)

**C.** Anti-MJ (anti-nuclear matrix protein 2)

**D.** Anti-melanoma differentiation-associated gene 5 (MDA5) (anti-CADM140)

**E.** Anti-histidyl-tRNA synthetase (anti-Jo1)

**Answers:**

**A.** Anti-HMGCR — Incorrect. Anti-HMGCR autoantibodies have been related to a subtype of immune-mediated necrotizing myopathies in patients with story of statin use.

**B.** Anti-p155/140 (anti-TIF$\gamma$) — Correct. Anti-p155/140 antibody is considered the most frequent myositis-specific antibody in JIIMs, present in 32% of cases.[4] Patients with CAJDM frequently have anti-p155/140 autoantibodies; these antibodies have been found in 75% of patients in some case series.[3] In adults, anti-p155/140 (anti-TIF$\gamma$) antibody is related to internal malignancy.

**C.** Anti-MJ (anti-nuclear matrix protein 2) — Incorrect. Anti-MJ autoantibodies are the second most frequent myositis-specific antibodies in JIIMs, present in 20% of patients.[4] Calcinosis is most common in patients with JIIMs with positive anti-MJ antibodies.

**D.** Anti-MDA5 (anti-CADM140) — Incorrect. Anti-MDA5 autoantibodies are rare in the Caucasian population with JIIMs. However, they are more frequently detected in East Asian patients, with a distinct clinical phenotype with digital necrosis and a rapidly progressive interstitial lung disease (ILD).

**E.** Anti-Jo1 — Incorrect. Antisynthetase autoantibodies (anti-Jo1, anti-threonyl-tRNA synthetase, anti-alanyl-tRNA synthetase, anti- glycyl tRNA-synthetase, anti-isoleucyl-tRNA synthetase, anti-asparaginyl-tRNA synthetase, anti-Ha, and anti-phenylalanyl-tRNA synthetase) are present in less than 5% of patients with JIIMs, with anti-Jo1 being the most frequent autoantibody.[4]

### Question 3: What is the main complication of CAJDM?

**A.** Internal malignancy

**B.** ILD

**C.** Calcinosis and vasculopathy

**D.** Renal involvement

**E.** Development of classic juvenile dermatomyositis (JDM)

**Answers:**

**A.** Internal malignancy — Incorrect. Internal malignancy has not been related to JDM, neither classic nor clinically amyophatic.[3,5]

**B.** ILD— Incorrect. Although it is considered extremely rare (<1%), there are some reports of ILD in children with CAJDM, especially related to anti-MDA5 autoantibody.[3]

**C.** Calcinosis and vasculopathy — Incorrect. Calcinosis and vasculopathy, mainly seen as intestinal perforation or skin ulcers, are considered the main complications of classic JDM. However, less than 5% of patients with CAJDM have been reported with these complications in different case series.[1,3,5]

**D.** Renal involvement — Incorrect. Renal manifestations could be present in idiopathic inflammatory myopathies. However, they are less common compared with other connective tissue disorders, including SLE and SSc.

**E.** Development of classic JDM − Correct. Fifteen percent to 25% of cases of CAJDM could evolve to classic JDM with clinical muscle disease during the first 2 years.[1] Therefore, follow-up is strictly recommended. To date, there is no reliable evidence of the benefit of early aggressive intervention to decrease the progression of more severe disease and muscle involvement.[5]

## Abbreviations used:

CAJDM: clinically amyopathic juvenile dermatomyositis
HMGCR: 3-hydroxy-3-methylglutaryl coenzyme A reductase
ILD: interstitial lung disease
JDM: juvenile dermatomyositis
JIIM: juvenile idiopathic inflammatory myopathy
MDA5: melanoma differentiation-associated gene 5
SLE: systemic lupus erythematosus
SSc: systemic sclerosis
TIF: transcription intermediary factor 1-gamma

**Conflicts of interest**

None disclosed.

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