Turkish Archives of Pediatrics

IMAGING OF PEDIATRICS
DOI: 10.5152/TurkArchPediatr.2023.23035

Clinically Amyopathic Juvenile Dermatomyositis

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Juvenile dermatomyositis (JDM) is the most common type of chronic inflammatory myopathy that presents with typical skin lesions such as Gottron papules, shawl sign, and heliotrope rash.[1] Clinically, amyopathic JDM is a subtype of dermatomyositis that can present with typical cutaneous manifestations but no muscle involvement, and diagnosis may be challenged in these patients due to lack of typical muscle weakness.[2] About 5%-20% of JDM patients present as amyopathic dermatomyositis.[3] Herein, we report an adolescent female patient who was diagnosed with clinically amyopathic JDM with typical skin manifestations related to JDM but without signs of muscle myopathy.

A 12-year-old female patient was referred to the pediatric rheumatology clinic with progressively worsening rashes with sun exposure on the face, neck, and extensor surfaces of the elbows, knees, and small joints of the hands for 7 years. Physical examination showed Gottron papules on the extensor surfaces of the elbows, knees, and small joints of the hands (Figure 1A), a shawl sign on the neck and shoulders (Figure 1B), and heliotrope rash on the face (Figure 1C), but muscle strength of both extremities was normal. In her family history, there was no history of rheumatologic disease and/or consanguinity between the parents. Laboratory workup revealed a positive antinuclear antibody (1:100 titer), a negative rheumatoid factor, normal levels of acute phase reactants, and normal levels of muscle enzymes, such as creatine phosphokinase, lactate dehydrogenase, aspartate transaminase, and alanine transaminase. In myositis-specific autoantibodies, anti-NXP2 antibodies were found as 2 positive titers. Eye examination, echocardiography, and lung imaging were normal. Subclinical myopathy was ruled out based on normal electromyographic and magnetic resonance imaging findings of the lower extremity. Skin biopsy from the extensor surface of the hand revealed an accumulation of mucin among collagen fibers, which is compatible with dermatomyositis. The patient was diagnosed with clinically amyopathic JDM due to typical skin manifestations related to JDM but no muscle involvement. Treatment with topical and systemic glucocorticoids, hydroxychloroquine,

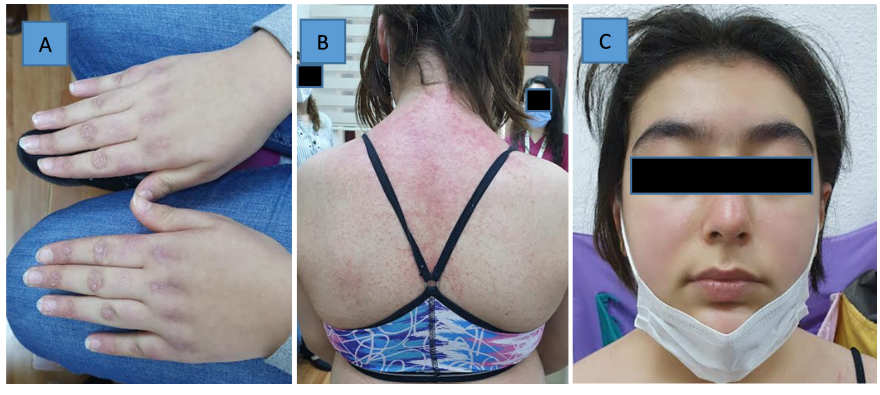

Figure 1. (A) Gottron papules on the extensor surfaces of the small joints of the hands. (B) Shawl sign on the posterior side of the neck and shoulders. (C) Heliotrope rash on the face.

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Received: February 16, 2023
Accepted: April 11, 2023
Publication Date: August 4, 2023

Cite this article as: Yıldırım DG, Bakkaloğlu SA. Clinically amyopathic juvenile dermatomyositis. *Turk Arch Pediatr.* 2023;58(5):553-554.

and methotrexate resulted in significant improvement in skin lesions at the follow-up visits.

There are rare case reports about clinically amyopathic JDM in childhood.[2,4] Interstitial lung disease is common, and corticosteroids (systemic and/or topical), hydroxychloroquine, and disease-modifying anti-rheumatic drugs are the main successful treatment options in clinically amyopathic JDM.[3] The typical cutaneous manifestations of JDM should be well known by clinicians because patients may not always present with muscle weakness as expected.

Informed Consent: A written informed consent obtained from the patient's parents.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – D.G.Y., S.A.B.; Design – D.G.Y., S.A.B.; Supervision – D.G.Y., S.A.B.; Materials – D.G.Y., S.A.B.; Writing Manuscript – D.G.Y., S.A.B.; Critical Review – D.G.Y., S.A.B.

Acknowledgments: The authors are grateful to the patient and her family.

Declaration of Interests: The authors have no conflict of interest to declare.

Funding: This study received no funding.

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