Open Journal of Clinical & Medical Images

www.jclinmedimages.org

Case Report

Open Access, Volume 2

Juvenile dermatomyositis in a child with calcinosis and joint ankylosis

Nour El Imene Ouni*; Nesrine Ben Salah; Amina Aounallah

Department of Dermatology, Farhat Hached University Hospital, University of Medicine, Sousse, Tunisia.

***Corresponding Author: Ouni Nour el Imene**
Department of Dermatology, Farhat Hached University Hospital, University of Medicine, Sousse, Tunisia.
Tel: +216-24041144;
Email: nourelimene286@gmail.com

Received: Sep 06, 2022
Accepted: Oct 21, 2022
Published: Oct 28, 2022
Archived: www.jclinmedimages.org

Introduction

Dermatomyositis is a multi-system autoimmune disorder primarily affecting the skin, skeletal muscle and blood vessels [1]. Among dermatomyositis, Juvenile Dermatomyositis (JDM) is rare [2]. It affects two to four children per million per year [1]. The average onset is in the 7th to 8th year of life, with a slight preference for the female gender [1]. Herein, we describe a case of JDM in a girl with calcinosis cutis and joint ankylosis.

Case report

A 13-year-old girl with a history of insidious onset and gradually progressive pain and weakness, predominantly affecting the proximal muscles of both upper and lower limbs, for the past 6 years, was admitted to our department. Physical examination revealed purplish heliotrope erythema of the eyelids, poikilodermatous reticulate erythematous lesions over the face and trunk and erythematous, flat papules and small plaques over metacarpophalangeal, proximal interphalangeal, and distal interphalangeal joints. Moreover, we noted tender and firm nodules over the knees and elbows. The patient had ankylosis of the elbows, hips and knees joints and she couldn't walk (Figure 1). She stopped all school and leisure activities The Laboratory investigations revealed normal serum-derived muscle enzymes (aldolase, creatine kinase, transaminase). Autoantibodies including ANA, anti-MI-2-, anti-Jo-1-, and anti-AMAM2 were negative. Electromyography was suggestive of a myopathic pattern. We retained the diagnosis of JDM based on the clinical presentation and the Electromyography. The patient was addressed to the department of physical medicine and rehabilitation. She has benefited from passive and then active physiotherapy exercises. During follow-up, the patient presented an increase in the range of all movements of the knee, elbow and hip joints.

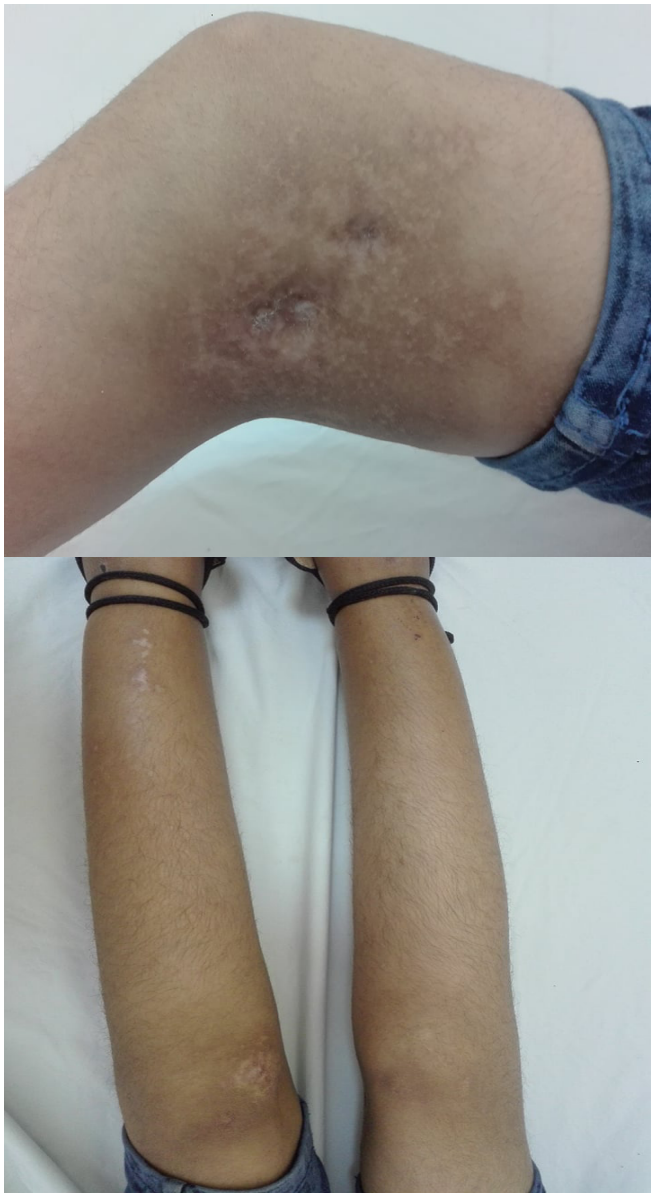

Figure 1A,B: Ankylosis of the knees joints.

Discussion

JDM is a rare systemic autoimmune vasculopathy characterized by muscle weakness and distinctive skin findings [1]. Its diagnostic criteria are currently still based on those established by Bohan and Peter in 1975, which include a characteristic skin eruption, proximal muscle weakness, elevated muscle enzymes, pathological muscle histology, and myopathic electromyographic changes [3]. JDM is diagnosed if four of these criteria are present, including the skin rash. The diagnosis is probable in the presence of two criteria in addition to the skin rash [3]. Our patient fulfilled the Bohan and Peter criteria for the diagnosis of probable JDM. The early diagnosis is often hampered by the nonspecific nature of the initial signs of JDM, such as fatigue, fever, weight loss, irritability, myalgia, and arthralgia. Identification of characteristic skin lesions may help establish an early diagnosis. Typical cutaneous lesions include a characteristic periorbital heliotrope rash, facial malar rash, Gottron papules and nailfold changes [1]. Muscle-derived enzymes are often elevated in JDM indicating muscle damage. In some cases, however, they may be normal, especially in the early and chronic stage of the disease and it has been shown that in untreated JDM after 4.7 months, the different muscle enzymes tended to be normal, which was the case in our patient [4]. Dystrophic calcification is one of the hallmark sequelae of JDM that occurs in 30%-70% of children with JDM [5]. It involves the deposition of insoluble calcium salts in the cutaneous and subcutaneous tissues. Patients may present with firm to hard irregular nodules or with larger subcutaneous plaques along the fascia or in the intramuscular connective tissue causing extensive areas of calcification, leading to functional impairment. Elbows, knees, buttocks, and shoulders are typically affected [5]. Although little is known about its pathophysiology, it is supposed that calcinosis may be mediated by activated macrophages and chronic local inflammation might be closely associated with its development [6]. Calcinosis has had a longstanding association with delay to diagnosis and initiation of therapy, but also occurs in chronic, severe disease [5,6]. Multiple treatment strategies have been tried including antiinflammatory drugs, drugs that affect calcium metabolism, and mechanical modalities. However, no therapy has proven to be reproducibly efficacious, and evidence in the literature is limited [5]. Although there are no established protocols for the management of JDM patients, there is significant evidence that suggests that early aggressive management improves outcomes, whereas delayed treatment is associated with poor outcomes and a higher rate of complications, which was, unfortunately, the case in our patient [7].

Conclusion

In conclusion, JDM is a rare but severe autoimmune vasculopathy that should be diagnosed and treated as early as possible to avoid disability and life-threatening complications.

Funding: None.

Conflict of interest: The authors declare that there are no conflicts of interest in this work.

References

1. Li D, Tansley SL. Juvenile Dermatomyositis-Clinical Phenotypes. Curr Rheumatol Rep. 2019; 21: 74.
2. Meyer A, Meyer N, Schaeffer M, et al. Incidence and prevalence of inflammatory myopathies: A systematic review. Rheumatology (Oxford). 2015; 54: 50-63.
3. Bohan A, Peter JB. Polymyositis and dermatomyositis (first of two parts). N Engl J Med. 1975; 292: 344-7.
4. Pachman LM, Abbott K, Sinacore JM, et al. Duration of illness is an important variable for untreated children with juvenile dermatomyositis. J Pediatr. 2006; 148: 247-53.
5. Hoeltzel MF, Oberle EJ, Robinson AB, Agarwal A, Rider LG. The presentation, assessment, pathogenesis, and treatment of calcinosis in juvenile dermatomyositis. Curr Rheumatol Rep. 2014; 16: 467.
6. Mukamel M, Horev G, Mimouni M. New insight into calcinosis of juvenile dermatomyositis: a study of composition and treatment. J Pediatr. 2001; 138: 763-6.
7. Kim S, El-Hallak M, Dedeoglu F, et al. Complete and sustained remission of juvenile dermatomyositis resulting from aggressive treatment. Arthritis Rheum. 2009; 60: 1825-1830.