Residência RP Pediátrica
Publicação Oficial da Sociedade Brasileira de Pediatria
ISSN-Online: 2236-6814

Submitted on: 02/28/2022
Approved on: 03/08/2022

INTERACTIVE CLINICAL CASE

A child with muscle weakness

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INTRODUCTION

Muscle weakness is present in the majority of myopathies and may occur in association with hypotonia. It may worsen slowly or remain clinically stable. Muscle weakness may also be present in patients with dystrophy, although differences relative to myopathy involve age of onset, affected muscles, and characteristic dystrophic alterations[1].

CASE REPORT

A seven-year-old male patient was taken to our service after experiencing myalgia in his legs and a progressive loss of the ability to walk associated with bouts of fever (temperature not measured) developing for about 20 days. A week prior to admission, purple, erythematous lesions appeared in the periorbital area and erythematous rash developed on the skin of the joints of the hands, elbows, and knees. His mother said he had no history of infection or comorbidity or a family history of autoimmune disease.

What is the most likely diagnosis?

Select the right statement concerning the patient's condition:

a) The main clinical manifestation is arthritis, characterized by pain, swelling and increased temperature in one or more joints.

b) The main manifestations include symmetrical proximal muscle weakness, elevated serum muscle enzymes, and skin lesions, of which heliotrope rash and Gottron papules are pathognomonic signs of the disease.

c) The first clinical manifestation is Raynaud's phenomenon, present in more than 90% of the patients and an important element in the early diagnosis of the disease in children.

d) Nephritis is a manifestation of concern that occurs in 50% of the patients.

DISCUSSION

Juvenile dermatomyositis (JDM) is a chronic autoimmune inflammatory myopathy that affects striated muscles, the skin, and other organs. The most common symptoms of JDM are proximal muscle weakness and pathognomonic skin findings, although some patients develop predominantly skin manifestations (hypomyopathic/amyopathic subtypes). * The disease is referred to as juvenile when it is diagnosed in individuals aged less than 16 years. Suggestive signs include heliotrope rash (purple, erythematous lesions with varying degrees of swelling in the periorbital area) and Gottron papules (maculopapular rash on the joints). The etiology of the condition is unclear, although it has been correlated with immune disorders and genetic predisposition for developing chronic vasculopathy[2]. Signs and symptoms vary and the Bohan and Peter criteria have been partly replaced, since electroneuromyography and muscle biopsy are no longer performed frequently due to invasiveness[3]. The EULAR/ACR criteria include four variables connected with muscle weakness and three related to skin manifestations, in addition to muscle enzyme levels (total creatine kinase and creatine kinase MB; aldolase; lactate dehydrogenase; and glutamic oxaloacetic transaminase). Patients are scored differently depending on whether a muscle biopsy has

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DOI: 10.25060/residpediatr-2022.v12n2-01

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Figure 1. Erythematous papules on joint surfaces of the hands.



Figure 2. Heliotrope rash and Gottron papules.

been performed. First line treatment comprises glucocorticoids often combined with methotrexate; immunoglobulin is used in cases of severe or refractory disease[4].

Please select the right option concerning the treatment of JDM:

I - General measures such as using sun screen, dieting, and proper hydration are of great relevance during patient follow-up.

II - Every patient diagnosed with JDM must be given glucocorticoids.

III - In general, patients with JDM progress well, particularly when they are diagnosed early on and treatment is promptly initiated and complied with.

a) Only statement I is correct.

b) Statements I and II are correct.

c) All statements are correct.

d) Only statements II and III are correct.

CONCLUSION

Recognizing the main signs and symptoms of JDM allows the early initiation of treatment, the prevention of sequelae, and the preservation of patient quality of life.

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