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CLINICAL CASE

Sarcoidosis in childhood. A rare systemic disease☆

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KEYWORDS
Childhood sarcoidosis;
Multiorgan involvement;
Hypercalcemia

Abstract
Background: Sarcoidosis is a systemic disease of unknown etiology that rarely occurs in children. It usually affects the lungs; however, it may involve various organs. It occasionally affects the general condition, and causes fever, hepatomegaly and splenomegaly.
Case report: We report the case of a twelve-year-old adolescent with late-onset childhood sarcoidosis which diagnosis was confirmed by lymph node histopathological study. The patient presented general condition, hypercalcemia, erythema nodosum, severe lung disorders, lymphadenopathy, hepatomegaly and testicular mass. He received treatment with steroids, with excellent clinical response.
Conclusions: We highlight the importance of considering the diagnosis of sarcoidosis in patients with hepatomegaly, lymphadenopathy, diffuse lung damage, erythema nodosum, testicular mass and hypercalcemia, as well as the need for a multidisciplinary approach to assess multiple organ involvement and the early beginning of steroid treatment in order to prevent the progression of the disease.

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PALABRAS CLAVE
Sarcoidosis infantil;
Compromiso multiorgánico;
Hipercalcemia

Sarcoidosis en la infancia. Una rara enfermedad sistémica

Resumen
Introducción. La sarcoidosis es una enfermedad sistémica de etiología desconocida que raramente se presenta en la infancia. Generalmente afecta los pulmones; sin embargo, puede involucrar diversos órganos. Ocasionalmente afecta el estado general, y origina fiebre, hepatomegalia y esplenomegalia.
Caso clínico. Se presenta el caso de un adolescente de doce años de edad con sarcoidosis infantil de inicio tardío, cuyo diagnóstico fue confirmado con un estudio histopatológico de ganglio linfático. El paciente cursó con afección general, hipercalcemia, eritema nodoso, alteraciones pulmonares graves, adenopatías, hepatomegalia y masa testicular. Recibió tratamiento con esteroides, con excelente respuesta clínica.
Conclusiones. Se resalta la importancia de considerar el diagnóstico de sarcoidosis en los pacientes con hepatomegalia, adenopatías, daño pulmonar difuso, eritema nodoso, masa testicular e hipercalcemia, así como la necesidad del abordaje multidisciplinario para valorar el compromiso orgánico múltiple y el inicio oportuno de la terapia con esteroides, con el fin de evitar la progresión de la enfermedad.

1. Introduction

Sarcoidosis is a chronic systemic disease of unknown etiology and worldwide distribution which is usually diagnosed in adults. Sarcoidosis in childhood is very rare.

Lungs are the most frequently affected organs; however, the disease can involve other organs such as eyes, skin, lymph nodes and joints. Less often it involves the nervous system, heart and urogenital tract, causing nephrolithiasis and a testicular mass; in some cases, fever of unknown origin with splenomegaly and hepatomegaly are observed.

The diagnosis of sarcoidosis is made by exclusion of other diseases. Therefore, once there is clinical suspicion, a biopsy of the organs involved shows, as a characteristic histopathological finding, the presence of noncaseating epithelioid granulomas.[1]

The first descriptions of sarcoidosis were made in Europe in the late nineteenth century. In 1877, in England, J. Hutchinson studied a patient with chronic skin lesions, arthritis and chronic renal failure, and named the skin findings *papillary psoriasis*. In France, in 1889, E. Besnier also described the skin lesions and named them *lupus pernio*. In 1899, in Denmark, C. Boeck labeled the skin histological lesions with the term *sarkoid* because of its similarity with sarcoma. In 1914, in Sweden, N. J. Schaumann described the systemic presentation of the disease; likewise, he pointed out that both Besnier´s *lupus pernio* and Boeck´s *sarkoid* were manifestations of the same disease, as the tissues affected in these patients showed granulomas which he called *benign lymphogranulomatosis* to differentiate them from *Hodgkin´s malignant granuloma*. The neurologic involvement in sarcoidosis was reported by C. Heerfordt in 1923 who described patients with *uveo-parotid fever* and lesion of cranial nerves.

The acute pulmonary form of sarcoidosis accompanied of mediastinal lymphadenopathy and erythema nodosum, arthritis and uveitis was described in Sweden in 1953 by S. Löfgren.

For more than 130 years, most of the studies on sarcoidosis have been performed in adults. However, pediatric cases have been reported since 1923. The condition was known as *Besnier-Boeck-Schaumann disease* until 1958 when the Sarcoidosis World Congress was carried out in London and the term *sarcoidosis* was generalized.[2,3]

Nowadays, accordingly to the international consensus established by the American Thoracic Society, the European Respiratory Society and the World Association for Sarcoidosis and Other Granulomatosis, sarcoidosis is considered to be a systemic granulomatous disease of unknown etiology, which usually affects adults, and is very rare in children.[4,5]

The incidence and severity of sarcoidosis vary in different regions of the world and in different ethnic groups probably due to variations in environmental exposures, the prevalence of HLA alleles and other genetic factors. Scandinavia, England, the United States and Japan have the highest prevalence of the disease; in Sweden, the morbidity rate in the general population is 64/100,000 and in the United States, 35/100,000.

There are very few epidemiological data on children. The rate of morbidity of childhood sarcoidosis is 0.29/100,000; however, this rate varies from 0.06/100,000 in children under 4 years to up to 1.02/100,000 in adolescents aged 14 to 15 years.[6-8]

Regarding mortality from sarcoidosis, during the period 1999-2010 the National Center for Health Statistics reported sarcoidosis as a cause of death in 10,348 of 29,176,040 deaths which represents a rate of 2.8/1,000,000 inhabitants. However, mortality in the African-American population was 12 times higher than in Caucasian population.[9]

In Mexico, the incidence of sarcoidosis is low, probably due to genetic factors or underreporting of cases. There-