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Case Report

Adult onset Still disease: A rare condition. Clinical experience with 17 cases☆

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ABSTRACT

Objective: To present current clinical experience and the clinical outcomes in the management of patients with adult Still disease.
Materials and methods: A retrospective study of a case series was conducted on of 17 cases diagnosed with adult Still disease during a period between 2009 and 2014 in 2 tertiary hospitals in Santiago de Cali.
Results: Of the 17 cases diagnosed and treated, it was found that 50% of patients had an elevated ferritin five times its normal value, being useful in the diagnosis. Less than 50% of cases were under 40 years, and about 50% of the population required combined treatment, with two patients requiring biological therapy to control their symptoms. Yamaguchi criteria were used for diagnostic correlation.
Conclusions: Adult Still's disease is a rare disease that requires a high index of suspicion, but it must be a disease to rule out, and it always requires management generally combined with schemes to improve the quality of life of patients.

Una patología poco frecuente: la enfermedad de Still del adulto. Experiencia clínica con 17 casos

Palabras clave:
Enfermedad de Still del adulto
Fiebre de origen desconocido
Serie de casos

RESUMEN

Objetivo: Dar a conocer la experiencia clínica en el mundo real y los resultados clínicos del manejo de los pacientes con enfermedad de Still del adulto.
Materiales y métodos: Se realiza un estudio retrospectivo de serie de casos, de 17 casos con diagnóstico de enfermedad de Still del adulto, en 2 clínicas de nivel 3 de Santiago de Cali, Colombia, en el periodo comprendido entre 2009 y 2014.

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Resultados: De los 17 casos diagnosticados y tratados se encontró que en el 50% de los pacientes había una elevación de ferritina de 5 veces su valor normal, siendo útil en el diagnóstico, menos del 50% de los casos fueron en menores de 40 años, alrededor del 50% de la población requirió tratamiento combinado y 2 pacientes requirieron terapia biológica para el control de sus síntomas. Se usaron los criterios de Yamaguchi para la correlación diagnóstica.

Conclusiones: La enfermedad de Still del adulto es una enfermedad rara que requiere alto índice de sospecha pero debe ser una enfermedad de descarte y, generalmente, siempre requiere manejo con esquemas combinados para mejorar la calidad de vida del paciente.

Introduction

Adult Still's disease (ASD) is an uncommon systemic disease of unknown etiology and inflammatory characteristics, with a broad spectrum of clinical presentations. There are no epidemiological data on its incidence or prevalence, and there is not a pathognomonic diagnostic test; given its behavior and the lack of diagnostic tests is considered a diagnosis by exclusion. Various criteria have been proposed for its diagnosis, including those of Yamaguchi that allow an accurate diagnostic approach.[1]

The pathophysiology of ASD is still not fully understood. Numerous causes such as infectious agents, genetic factors and alteration in the regulation of the immune response and the apoptosis have been postulated, but there are still not concrete data.[2–6] The postulated infectious agents, based on temporal relationships between the onset of the disease and the serological evidence, are parvovirus B19, rubella, echovirus 7, Epstein–Barr, cytomegalovirus, *Coxsackie* B4, *Chlamydia* and *Yersinia*; however, it has not been possible to establish a definitive relationship.[2]

The immunological pathways of the disease have been considered the most significant in the pathophysiology. The studies have demonstrated that the proinflammatory interleukins (IL) play a vital role in the pathogenesis[7,8] and elevated levels of IL-1 have been detected in the untreated disease, which has been correlated with the disease activity and severity.[9] Likewise, this acute phase response is considered the responsible for the stimulation of the synthesis of ferritin and, particularly, the IL-18 favors such stimulation and plays an important role in the manifestations of the disease, by correlating with the levels of neutrophils in blood.[10]

The main manifestations of the disease are intermittent fever predominantly in the evenings, usually for less than 4 h, associated with evanescent salmon colored maculopapular rash on the trunk and extremities. It can be accompanied by joint commitment that involves mainly the wrists, knees and ankles, although whichever joint may be affected, it also tends to be symmetrical and bilateral. In addition, it may occur odynophagia, hepatosplenomegaly and serious manifestations such as pleuropericarditis, pleural effusion and interstitial pneumonitis.[11,12] Paraclinically, it may occur with leukocytosis and neutrophilia in more than 80% of cases, alterations of liver function tests, elevation of ferritin up to 5 times the normal upper value and elevation of glycosylated ferritin. The blood cultures and autoimmunity tests are negative.[13] Several classification criteria had been used, being those of Yamaguchi the most commonly used given their sensitivity (93%). Other recognized criteria are those of Cush and Fautrel with a sensitivity of 80% for both.[11]

A series of 17 cases diagnosed in the city of Santiago de Cali, in 2 centers of level 3 of the city, is presented. The clinical description of the patients and the diagnostic method is made, and is also described how was the treatment of the patients and the results previously obtained. The lack of evidence, due to the low prevalence of the disease, motivates the realization of this type of case series for a better understanding of the behavior of the disease in our population.

Materials and methods

Retrospective study of a case series. 17 cases with diagnosis of ASD based on Yamaguchi's criteria, in 2 clinics of level 3 of Santiago de Cali, in the period between 2009 and 2014 (15 from the Imbanaco Medical Center and 2 from the Comfenalco Valle Unilibre Corporation). The age, the gender, the clinical classification, the time elapsed between the onset of the symptoms and the diagnosis, the treatment used, and the clinical and laboratory characteristics are analyzed. Only the percentages were used for the analyses (Table 1).

Results

Clinical characteristics of the patients

The curse of the disease had an approximated duration of 40 days between the onset of the symptoms and the diagnosis. The cases of rapid diagnosis were linked to a more timely consultation to a rheumatologist. 95% of patients had fever, all since the beginning of the disease. All patients had the characteristic skin rash. 86% of patients presented joint involvement at some point of the disease, generally associated with the febrile episodes. Half of the patients had odynophagia and 30% of the total had palpable adenopathies on the physical examination. Only one patient had hepatomegaly with transient alteration of the liver function tests (Fig. 1).