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Still's disease in adolescence

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Introduction: Still's disease is a rare systemic inflammatory disease of unknown etiology and a frequent cause of fever of unknown origin in adults. It affects both sexes in the same proportion, and in 75% of cases is between 16 and 35 years old.1 Juvenile systemic idiopathic arthritis, also known as Still's Disease, is characterized by high fever spikes, which persist for more than two weeks, often associated with a non-fixed, evanescent erythematous rash. It is called "Juvenile" because it typically affects children under the age of 16. By systemic, it is said that along with joint inflammation it usually begins with symptoms and signs of systemic (body-wide) illness, such as elevated fevers, swelling of the gland, and involvement of the internal organ. By idiopathic it is meant that there is no known cause for the disease. Abdominal pain and myalgia can intensify during peak fever. Other systemic features include hepatosplenomegaly, generalized lymphadenopathy, and serositis (pleuritis or pericarditis).2This inflammation can wreck affected joints, particularly wrists. Treatment involves medicines which help control inflammation, such as prednisone. The signs and symptoms of this disorder that resemble those of other disorders, including lupus, and a form of lymphoma called cancer.

According to the International League of Associations for Rheumatology (ILAR)3, JIA is classified into seven categories that include the full spectrum of clinical manifestations of this entity: systemic arthritis, oligoarthritis, polyarthritis with negative rheumatoid factor, polyarthritis with positive rheumatoid factor, psoriatic arthritis, enthesitis-associated arthritis, and unclassified arthritis. Systemic Juvenile Idiopathic Arthritis is the most severe form and has been recognized as a unique form in chronic childhood arthritis, both for its clinical and epidemiological characteristics. It is an entity of difficult diagnosis due to its nonspecific symptoms and shared by several common pathologies in pediatric ages.4

Case Report: JPLB, male, 14 years old, brown was admitted in the pediatrics department. Young man previously healthy, history of fever, polyarthralgia in hands, wrists, feet and ankles with wrist arthritis for about 10 days, with sporadic rash. No report of previous infections. Lucid and oriented patient, acyanotic, anicteric, flushed, hydrated, average of 3 febrile peaks per day (38ºC). Hemodynamically stable. Normal physical exam. Laboratory exams: Hb = 12.0; Hto = 37.3; Leukogram = 26,470; Rod = 11%; Segmented = 84%; PCR = 178; Cr = 0.4; Urea = 20; TGO = 28 / TGP = 24; CPK = 465; Rheumatoid Factor = Negative; Ferritin = 2,075.

Serology was performed for chikungunya less than 10 days after febrile episode (IgM and IgG = negative) and hemogram (CBC) with leukocytosis and elevated ferritin. Thus, the diagnosis of systemic juvenile idiopathic arthritis was made - compatible clinical and laboratory picture. It was started treatment with prednisone 20 mg / day and Methotrexate. The patient evolved with improvement of symptoms and laboratory improvement: Hb = 15.1; Hto = 45.3; Leukogram = 5,300; CRP = 0.2; ESR = 02 and Ferritin = 20.8. The patient was discharged one week after hospitalization and he was referred for Rheumatology outpatient clinic follow-up.

Discussion: Systemic Juvenile Idiopathic Arthritis (SJIA) is one of the leading causes of short- and long-term disability, affecting approximately 250,000 children in the United States.5 It is usually characterized by high fever, evanescent rash and arthritis, in addition to multisystem involvement, renal and neurological involvement are rare. Serum ferritin levels above 1,000 ng /ml correlate with disease activity.1

The clinical course of systemic juvenile idiopathic arthritis is highly variable. In about half of the patients, the disease is monocyclic, or characterized by relapses followed by intervals of remission. However, when systemic characteristics are controlled, arthritis usually subsides. The long-term prognosis for these patients is generally good. In the other half of the patients, the disease follows a continuous course. In many cases, systemic symptoms eventually resolve, with chronic arthritis being the main long-term problem.2

Immunological evidence includes abnormalities in natural killer T lymphocytes (NK), caused by reduced expression of genes involved in innate immunity. Therefore, an aberrant phagocyte activation occurs that ends in the secretion of pro-inflammatory cytokines Interleukin 1 (IL-1), Interleukin 6 (IL-6) and Interleukin 18 (IL-18), as well as pro-inflammatory proteins such as calcium-binding proteins.6 A strong association with macrophage activation syndrome is an important feature in systemic juvenile idiopathic arthritis.7 The established treatment for SJIA is with non-steroidal anti-inflammatory drugs and systemic corticosteroids. Methotrexate is an alternative in the treatment.8

Conclusion: The diagnosis of Still's disease is difficult. The etiology is still unknown, it is a multifactorial pathology influenced by environmental and genetic factors. The diagnosis is essentially clinical and there is no laboratory test that can confirm it, so there are numerous differential diagnoses. But it should be considered in cases of fever of undetermined origin in adolescents, after excluding infectious and neoplastic causes.

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