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ORIGINAL ARTICLE

Atypical Kawasaki disease associated to cholestatic syndrome: case report

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Abstract

Objectives: To report the case of a child with atipical Kawasaki disease associated with cholestatic syndrome. Kawasaki disease is an inflammatory syndrome that affects small and medium-sized vases, which cause is unknown. The diagnostic is clinical and based in classic criterions, which are commonly reported symptoms, does not including cholestatic jaundice nor hepatomegaly because they are un-usual findings. **Case Report:** 6-year-old female presented persistent high fever, rash and gastrointes-tinal symptoms. She developed with conjunctival injection, strawberry tongue, and cracked lips, with-out improvement of the fever after the empiric antibiotic treatment. In elapsing of the disease, the rash got improvement and appeared cholestatic syndrome and classic symptoms of the disease of complete Kawasaki (unilateral cervical lymphadenopaty, conjunctival injection, and hand desquama-tion), that it made possible her diagnosis and treatment, with improvement of cholestatic syndrome, without aneurysm appearance. **Conclusions:** This paper shows the importance of including Kawasaki disease as differential diagnosis in cases of jaundice feverish syndromes, allowing their treatment in skilled time end prevent complications.

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INTRODUCTION

Kawasaki disease (KD) is a multisystem inflammatory syndrome, of unknown etiology and variable clinical expression.[1] Characterized by small and medium vessel vasculitis, its main complication is coronary artery aneurysm[2]. Its diagnosis is based on clinical criteria, as there are no tests or laboratory findings specific to the disease.[2] However, some patients present with atypical conditions, which often leads to a delay in diagnosis and treatment.[2] The involvement of the gastrointestinal tract, with diffuse abdominal pain and cholestatic syndrome has rarely been described in KD.

The present study reports the case of a female child with an atypical presentation of the disease and aims to reinforce the need for knowledge about clinical manifestations not covered by the classical criteria.

CASE REPORT

A 6-year-and-8-month-old female patient, previously healthy, sought emergency care with a complaint of persistent fever (up to 39°C), associated with generalized and pruritic maculopapular rash, diffuse abdominal pain, especially postprandial nausea and vomiting of food content. Admitted to a pediatric hospital, she evolved on the second day of the disease with bilateral conjunctival hyperemia, raspberry tongue and cleft lip. She received empirical treatment with ceftriaxone and symptomatic medication, without improvement of the febrile condition.

On the 8th day of the illness, the child had an improvement in the exanthema, but evolved with jaundice, choluria and fecal acholia, and she was transferred to a tertiary pediatric hospital.

Upon admission, she had had a daily fever for 11 days and had furfuraceous desquamation on her face and trunk, and lamellar on her hands, without arthritis, oropharynx without alterations. In her sclera, in addition to jaundice (2+/4+), he had mild residual hyperemia. She also had a palpable ganglion in the right posterior cervical chain, measuring approximately 1.5 cm in diameter, with a fibroelastic and painless consistency. Upon abdominal palpation, she had a liver 4 cm from the right costal margin, painful and with a regular surface.

Based on the clinical history and physical examination, Kawasaki disease with cholestasis was diagnosed and treatment was immediately started with intravenous Human Immunoglobulin, 2g/kg in a single dose, associated with acetylsalicylic acid (ASA) at a dose of 80 mg/kg/day.

Her admission exams showed Hb: 10.6 mg/dL; Ht: 31.9%; VCM: 78.6 fl; HCM 26.1 pg; Leukocytes: 19,500/mm³ (segmented 80%, lymphocytes 12%, rods 0.5%, eosinophils 6.3%); Platelets 677,000/mm³; CRP 18.1mg/dL (reference value < 6); 120mm VHS; TGO: 102 U/L; TGP: 119 U/L; Alkaline Phosphatase: 1593 U/L; GT range: 576 U/L. Serology for Cytomegalovirus, Toxoplasmosis, Hepatitis A, B and C, Epstein Baar Virus and Dengue, negative Widal reaction and Plasmodium test. Anti-nucleus antibody, anti-smooth muscle and anti-mitochondria were negative, excluding the diagnosis of autoimmune hepatitis. Unaltered total abdominal ultrasound.

The patient evolved with remission of fever, improvement in abdominal pain and gradual reduction of cholestatic signs 24 hours after the start of treatment.

An echocardiogram performed at the time of diagnosis showed no changes in the coronary arteries and the patient was discharged from hospital 72 hours after the start of treatment, using ASA at a dose of 5mg/kg/day, for outpatient follow-up in a Pediatric Rheumatology unit. Subsequent echocardiograms also showed no involvement of coronary arteries.

DISCUSSION

KD is a self-limited vasculitis, of unknown origin, whose diagnosis is based on clinical criteria: high fever (from 38° to 40°C), for at least five days, associated with at least four out of five of the following signs: 1. Polymorph exanthema (without vesicles); 2. Bilateral conjunctival hyperemia without exudate; 3. Changes in lips and oral cavity, such as enanthema, fissures, raspberry tongue, diffuse edema of the oral mucosa and pharynx; 4. Cervical lymphadenopathy greater than 1.5 cm in diameter and usually unilateral; 5. Changes in the limbs: initially erythema of the palmar and plantar regions, followed by edema of the hands and feet, and in the subacute phase, there was evidence of periungual lamellar desquamation.[3]

In the case reported here, the patient had fever for more than 5 days associated with five of the following criteria: rash, unilateral cervical adenomegaly, conjunctival hyperemia, characteristic lesions of the oral cavity and changes in the extremities observed on physical examination. In this case, however, we highlight the development of cholestatic condition in the course of the disease, with jaundice, which led to a delay in the diagnosis and specific treatment in the recommended time.

Although gastrointestinal symptoms such as nausea, vomiting, diarrhea and abdominal pain are frequently observed in patients with KD,4 cholestatic jaundice and hepatomegaly, sometimes with associated splenomegaly, are reported in the literature as uncommon findings. Interestingly, like the case described, they usually occur in the absence of cardiac alterations.[1,5].

Few studies have reported jaundice as a prominent sign in the clinical picture of KD. In most of them, the patients initially presented with febrile cholestatic syndrome, later fulfilling the criteria established for KD[6-9].Taddio et al.[10] also emphasize this atypical initial presentation, reporting 5 cases, of which only two they had other clinical alterations of the disease at the time of hospital admission. Koca et al.[11] reported a case in which the definitive diagnosis was established only after evidence of coronary lesion, on the fourteenth day of the disease. Our patient presented cholestatic symptoms

during KD and, despite this, cholestasis was a determining factor for the delay in diagnosis, probably due to the rarity of such manifestations and frequent differential diagnoses in our region, especially the contagious diseases.

Other signs not covered by the classical criteria, but which are relatively common, include arthritis, irritability in response to aseptic meningitis, pneumonitis, gastroenteritis, colitis, dysuria and otitis. In addition to them, the following are also rare: cranial nerve palsy, renal involvement, gallbladder hydrops, gastrointestinal ischemia, macrophage activation syndrome and inappropriate antidiuretic hormone syndrome.[1,5] Such alterations corroborate the diagnosis of atypical cases or incomplete KD, where the patients do not meet all the proposed criteria.

Due to the existence of other more frequent causes of icteric-febrile syndrome in our country, an investigation was carried out for viral and autoimmune hepatitis, as well as for other differential diagnoses, including those peculiar to our region with a tropical climate, in an underdeveloped country . These surveys were negative. Abdominal ultrasound, in the context of the investigation of liver status, also had a normal result, and it is important to emphasize that some references point to occasionally found alterations, such as hydrops or thickening of the gallbladder wall, liver enlargement and of bile duct ectasia[1,10-12].

Although not part of the diagnostic criteria, KD is associated with a high incidence of coronary artery aneurysm in untreated patients, and this complication is the main cause of morbidity and mortality, reinforcing the need for diagnosis and treatment until the tenth day of the disease.[5] The investigation with an echocardiogram in our patient showed no alterations, both at the time of diagnosis and during outpatient follow-up. It is known that timely treatment significantly reduces the risk of this serious complication.[3]

Therefore, in a typical or highly suspicious condition, treatment should be promptly initiated with acetylsalicylic acid (80-100mg/kg/day) and Human Intravenous Immunoglobulin (at a dose of 2g/kg in a single dose).[3] We received the patient on the eleventh day of illness, but the treatment was justified by the marked presence of inflammatory signs, such as persistent fever, cholestasis and increased evidence of the acute phase. In fact, there was a therapeutic response after the start of treatment, as there was a resolution of the febrile condition and regression of the signs of cholestasis. In addition, there was a favorable evolution, as the patient did not present aneurysms.

In this case report we stress that even in a tropical climate region, where infections are the main cause of febrile--jaundice syndrome, we should include Kawasaki's disease as a differential diagnosis, since its specific treatment in a timely manner prevents serious complications of early installation.

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