

Case 9

Caso 9

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Figure 1 - Patient at the peak of infection, showing confluent erythematous papules on the trunk and limbs, characterizing morbilliform exanthem.



Figure 2 - Notice the presence of palmar exanthem and edema.

A 12-month old male infant presented with high fever for 11 days with partial response to antipyretics. On the fourth day, he was taken to emergency care, where he was diagnosed with infection of the upper airways and submitted to antibiotic therapy. A polymorphic rash was soon found on his torso and then limbs, and the fever did not improve. Upon his return to care, he was diagnosed with urticaria and treated with antihistamines, with no improvement. Seeking medical care for the third time, in addition to a body temperature of 39°C and the rash, palmar and con-

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conjunctival bilateral hyperemia and labial, lingual, and oropharyngeal erythema were also found.



Figure 3 - Patient in convalescence, with the back of the hands showing periungual desquamation.



Figure 4 - Notice erythema and exanthem on lips.

Based on clinical history and the images, which is the most likely diagnosis?

- measles,
- Kawasaki Disease,
- scarlet Fever,
- systemic Juvenile idiopathic arthritis (Still's disease).

DIAGNOSIS

Persistent fever for five or more days, accompanied by four out of five of the following clinical

criteria, strongly suggest the diagnosis of Kawasaki Disease (KD):

- non-purulent conjunctivitis,
- strawberry tongue, oropharyngeal erythema and edema, with labial fissures and erythema,
- erythema and edema of the hands and feet, which develops into periungual desquamation,
- scarlatiniform, morbilliform, or polymorphic rash, appearing first in the torso and then in limbs,
- cervical lymphadenomegaly.

In measles the maculopapular rash appears first behind the ears and spreads to forehead, neck and torso, with no edge desquamation. Whitish spots near the molars teeth (Koplik spots) are pathognomonic for the disease and precede the rash. It is followed by diarrhea and respiratory symptoms. Measles has nowadays been eradicated in Brazil by vaccination.

Scarlet fever presents with pharyngitis and tonsillitis, with coating and maculopapular exanthem similar to sandpaper, more pronounced in the creases and associated with perioral pallor (Pastia's and Filatov's signs). It responds quickly to therapy with antibiotics and is rare in infants, having higher incidence in children over five years.

Systemic Juvenile idiopathic arthritis (Still's disease) manifests mainly with fever, exanthem in the torso and limbs, arthralgia, arthritis and odynophagia. It is not associated with conjunctivitis, tongue and oropharynx erythema, or edge desquamation. Exanthems are often more pronounced during fever peaks.

CASE DISCUSSION

Kawasaki Disease (KD) is an acute multisystem self-limited vasculitis that affects vessels of small and medium caliber. The heart is the main affected organ, although others, such as lungs, intestines and the central nervous system may also be compromised. Strictures and aneurysms in coronary arteries are the main heart complications and may occur in up to 25% of untreated patients.

KD has a high incidence among children up to five years of age, and is rare before six months or after age eight. Slightly more frequent in boys, it is the leading cause of acquired heart disease in children in the United States and Japan. Its etiology remains unknown but KD is believed to be triggered by a still unidentified infectious agent that causes clinical changes in genetically susceptible individuals.

The diagnosis is essentially clinical, based on the aforementioned signs and symptoms, but can be uncertain due to similarities with other clinical conditions. Some laboratory findings can reinforce the clinical hypothesis, including left-deviating leukocytosis and increased ESR and C-reactive protein. There may also be normocytic, normochromic anemia, thrombocytosis and sterile pyuria.

Cardiovascular monitoring is essential after KD is diagnosed. Initial examination includes two-dimensional echocardiography with high sensitivity and specificity to detect abnormalities in the proximal segments of the coronary arteries. Other examinations may be performed based on specific indications, such as nuclear magnetic resonance imaging, computerized tomography and angiography.

Treatment requires hospitalization for administering intravenous immunoglobulin (IVIG) for the first 10 days of diagnosis so as to reduce the risk of coronary complications and shorten the duration of symptoms. Associated use of acetylsalicylic acid is also recommended due to its anti-inflammatory and antiplatelet effects.

Recurrence of KD is rare. Due to the likely increased risk of morbidity in adults with the preexisting disease, all children should be submitted to follow up exams every 3-5 years.

RELEVANT ASPECTS

- Kawasaki Disease (KD) is an acute multisystem self-limited vasculitis that affects vessels of small and medium caliber,
- it has high incidence before the age of five years,
- its diagnosis is essentially clinic and uncertain,
- heart involvement occurs in 25% of the non-treated patients, with aneurysm formation and coronary strictures,
- after diagnosis, monitoring cardiovascular function is essential, and echocardiography is the method of first choice,
- treatment with intravenous immunoglobulin (IVIG) and acetylsalicylic acid initiated on the first 10 days of the disease reduces the chances of complications.

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