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DIAGNOSTIC CRITERIA FOR KAWASAKI DISEASE: A LITERATURE REVIEW

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Abstract: Kawasaki disease manifests as systemic vasculitis that mainly affects medium-sized vessels, particularly the coronary arteries. It is a rare disease that mainly affects children, and is almost exclusively a pathology restricted to childhood. Kawasaki disease, despite being self-limited, can lead to important and potentially fatal complications if it is not identified in a timely manner.
Therefore, it is necessary to elucidate the diagnostic methods that are associated with early identification of the clinical condition, using already established criteria, aiming to reduce such complications and seeking a significant reduction in infant morbidity and mortalityl.
Keywords: Kawasaki disease. Diagnosis. Mococutaneous Lymph Node Syndrome. Complications.

INTRODUCTION

Kawasaki disease, also known as Mucocutaneous Lymph Node Syndrome, is a systemic, self-limited vasculitis. It causes fever and other acute inflammatory manifestations, lasting an average of 12 days if left untreated. It is characterized by affecting medium vessels, especially the coronary arteries, and can cause aneurysms or dilations1. Initially described in Japan by Tomisaki Kawasaki, this pathology is more prevalent in childhood, in children under 5 years of age, more frequently in East Asian countries, especially in Japan. It is the most common cause of heart disease acquired in childhood, in developed countries, leading to complications in the coronary arteries in around 25% of cases, with aneurysm being the main involvement. Furthermore, cardiomyopathies with depressed myocardial contractility, heart failure, myocardial infarction, arrhythmias and peripheral arterial occlusion are reported in the literature, causing significant morbidity and mortality.

Although it is more common among children of Japanese descent, with an incidence of approximately 112 cases per year per 100,000 children under the age of five, it can occur in children from any continent and even in adults2. However, the etiology of Kawasaki disease has not yet been well elucidated, although there is evidence of an infectious cause, but autoimmune factors may be involved. Furthermore, the diagnosis is essentially clinical, therefore using criteria developed by the Japanese Kawasaki Disease Research Committee or the American Heart Association (AHA), which are extremely important3. Several studies demonstrate that early diagnosis is essential to achieve the ideal result, both from treatment and the potential reduction in morbidity and mortality.

OBJECTIVES

The present study's primary objective is to elucidate the main diagnostic forms of Kawasaki disease, based on the literature, aiming at early recognition of the disease and better prognosis, with a focus on reducing morbidity and mortality.

METHODOLOGY

The research was carried out in the first half of 2023, of a descriptive type, using the literature review model. A careful bibliographic search was carried out on the topic, in the SCIELO and PubMed databases, using the descriptors "Kawasaki Disease" and "Diagnosis", with the Boolean operator "AND" as a connector of the words. The inclusion criteria cover: (a) articles available free of charge, (b) in English and Portuguese and (c) without year restrictions, which address Kawasaki disease and its diagnosis. However, articles that did not address the topic and those with repeated information were excluded. After choosing the criteria, the abstracts classified as included were read in full, regarding the gaps addressed in the research.

RESULTS

The diagnosis of Kawasaki Disease (KD) is clinical, although there are no clinical characteristics that are pathognomonic of the disease. Isolated laboratory and imaging tests also do not confirm the diagnosis, but they can help the doctor during the investigation to exclude differential diagnoses. For diagnosis, therefore, history and physical examination become extremely important.

The diagnostic criteria developed by the Japanese Kawasaki Disease Research Committee (Table 1) are more used in Asian countries, while in the West the criteria developed by the American Heart Association (AHA)4 are more used. Both are based on clinical findings, but the main difference between them is the need for the presence of fever for more than 5 days as a prerequisite5,6 and the description of the size of the cervical lymph nodes6. According to the AHA, the diagnosis is made based on the mandatory criterion of high and persistent fever for at least 5 days, associated with at least 4 of the 5 clinical criteria: Changes in the lips and oral cavity, conjunctival hyperemia, lymphadenopathy, diffuse and polymorphic rash, and changes in extremities (Table 2).

Fever for more than 5 days
Bilateral conjunctivitis
Changes in lips and oral cavity
Polymorphic rash
Extremity changes
Acute non-purulent cervical lymphadenopathy

Table 1: Japanese Kawasaki Disease Research Committee criteria for diagnosing Kawasaki disease - At least five of six criteria

Mandatory criteria: Persistent fever for at least 5 days, plus 4 of the following symptoms:
Changes in lips and oral cavity - Erythema and fissure on the lips and/or hyperemia in the oropharyngeal mucosa, and/or prominence of the lingual papillae ("franberry tongue")
Conjunctival hyperemia - Non-exudative bilateral bulbar conjunctivitis.
Lymphadenopathy - cervical, ≥5 cm in diameter, usually unilateral
Diffuse and polymorphic rash
Extremity changes - Palmar and/or plantar erythema and edema of hands and feet and/or periungual peeling in the subacute phase

Table 2: Diagnostic criteria for Kawasaki Disease, according to the American Heart Association

Fever is high (>39°C) and remitting, persisting for approximately 1 to 3 weeks without treatment[4,7]. After the onset of fever, conjunctival hyperemia appears, characterized by bilateral, non-exudative bulbar conjunctivitis, sparing the limbic region. Oral changes are observed including erythema, fissures, scaling and bleeding on the lips and "strawberry tongue" (erythema and prominence of the fungiform papillae) and erythema of the oropharyngeal mucosa. Cervical lymphadenopathy is, in most cases, unilateral and ≥ 5 cm in diameter. The erythematous skin rash usually appears about 5 days after the onset of fever and is polymorphous, with a maculopapular shape, and scarlatiniform erythroderma and erythema multiforme are also common[7]. The exanthema is diffuse, affecting the trunk, limbs and groin, with early desquamation.

Furthermore, changes in the extremities can present themselves in different ways, often with edema and hard erythema on the soles of the feet and palms of the hands, which is painful in an acute condition. 2 to 3 weeks after the onset of fever, during the subacute phase, desquamation of the fingers and toes may occur, progressing to the palms and soles.

Some findings are common in laboratory

tests in these patients, even though they are not specific, which can help in the diagnosis. It is common to present in the hematological examination, leukocytosis with neutrophilia (above 15,000/mm3), thrombocytosis (greater than 450,00/mm3), normocytic and normochromic anemia, elevation of inflammatory activity tests, such as ESR and CRP, and biochemical changes such as elevation of transaminases, albumin greater than 3 g/dl and hyponatremia. Urinalysis may show sterile pyuria; CSF analysis may show moderate sterile pleocytosis in case of aseptic meningitis.

In some cases, there are children who do not develop all the signs and do not meet the typical AHA criteria, which is known as incomplete Kawasaki Disease. This form is more prevalent in infants, especially those under 6 months of age, and classic symptoms, such as cervical lymphadenopathy and changes in the extremities, may not be present, and persistent fever may be the only clinical finding. The hypothesis of the incomplete form is raised, in patients with fever lasting more than 5 days, with only 2 or 3 of the main clinical criteria. The laboratory findings already mentioned above can support the diagnosis of atypical Kawasaki disease and the echocardiogram also plays an important role in the diagnosis, as it is possible to find cardiac abnormalities that, despite not being part of the diagnostic criteria for the typical form, reinforce in uncertain cases or when an atypical disease is suspected. For this reason, in cases of infants with fever for at least 7 days, laboratory tests and an echocardiogram must be performed to aid in the diagnosis.

CONCLUSION

Based on the scientific evidence collected, it is possible to affirm that knowledge about anamnesis and physical examination are the necessary tools for health professionals to recognize the characteristic changes of Kawasaki Disease. This way, it is possible to provide an early diagnosis and a better prognosis for the patient. Furthermore, the impact of socio-educational measures on health also needs to be highlighted, after all, raising awareness about this disease and its symptoms allows people who are not in the health sector to identify and seek treatment. This knowledge can be passed on in different ways. Firstly, family health units must be highlighted, which can organize public lectures and distribute pamphlets that, in accessible language, transmit to the population the information necessary to understand this disease. Furthermore, a partnership between the Ministry of Health and open television channels could be a great instrument to disseminate such information at times with a large number of viewers. These actions will allow democratic access to health, uniformly reaching different social classes.

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