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KAWASAKI DISEASE

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Abstract:

Kawasaki disease is a disease in which blood vessels throughout the body become inflamed. The most common symptoms include a fever that lasts for more than five days and is controlled by usual medications, large lymph nodes in the neck, a rash in the genital area, and red eyes, lips palms or bottom of the feet. The cause is unknown. It may be due an infection triggereing an autoimmune response in those who are genetically predisposed. It is not spread between people. Diagnosis is usually based on a person's signs and symptoms. Other tests such as an ultrasound of the heart and blood tests may support the diagnosis. Other conditions that may present similarly include scarlet fever and juvenile rheumatoid arthritis. Initially treatment is typically with high doses of aspirin and immunoglobulin. Usually with treatment fever resolves within 24 hours and there is a full recovery. If the coronary arteries are involved ongoing treatment or surgery may occasionally be required. Without treatment coronary artery problems occur in up to 25% and about 1% die. With treatment the risk of death is 0.17%.

Key Words: Inflamed, Triggering & Lymph Nodes

1. Introduction:

Kawasaki per is a rare. Disease that affects between 8 and 67 per 1000,000 people under the age of five except in Japan where it affects 124 per 100,000. It is much less common after the age of five. It is much less common after the age of five. Boys are more commonly affected than girls.

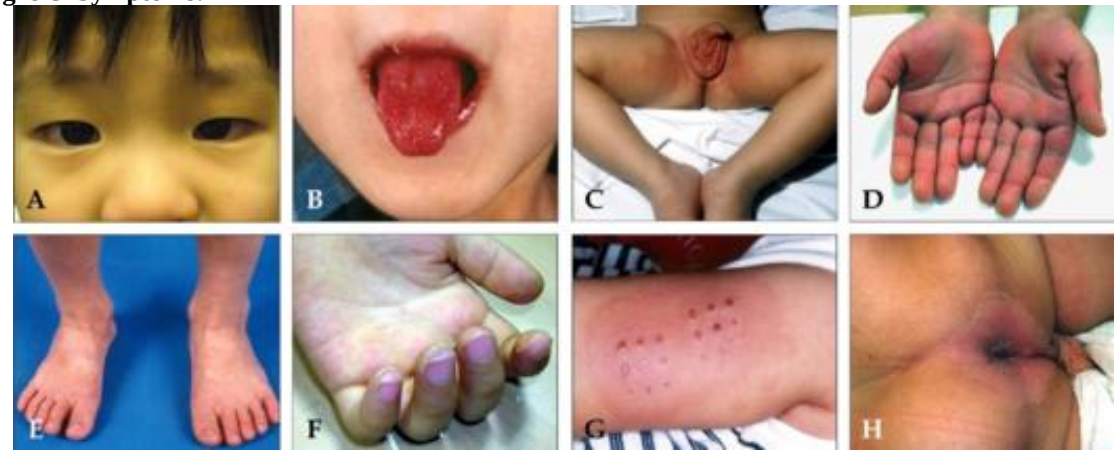
2. Definition:

Kawasaki disease is a disease in which blood vessels throughout the body become inflamed. The most common symptoms include a fever that lasts for more than five days and is controlled by usual medications, large lymph nodes i the neck, a rash in the genital area, and red eyes, lips palms or bottom of the feet.

3. Cause and Risk Factor:

As the cause(s) of Kawasaki disease remain unknown, the illness is more accurately referred to as Kawasaki syndrome. Its cause is widely hypothesized to involve the interaction of genetic and environmental factors, possibly including an infection in combination with genetic predisposition to an autoimmune mechanism. The specific cause is unknown, but current theories center primarily on immunological causes. Evidence increasingly points to an infectious cause,[92] but debate continues on whether the cause is a conventional antigenic substance or a super antigen. Researchers at Boston Children's Hospital reported, "some studies have found associations between the occurrence of Kawasaki disease and recent exposure to carpet cleaning or residence near a body of stagnant water; however, cause and effect have not been established."

4 Signs & Symptoms:



The most common symptoms include a fever that lasts for more than five days. Bilateral conjunctival inflammation was reported to be the most common symptom after fever. It typically involves the bulbar conjunctivae, is not accompanied by suppuration, and is not painful. It usually begins shortly after the onset of fever during the acute stage of the disease may be present on slit-lamp examination. Iritis can occur, too. Kawasaki disease presents with set of mouth symptoms, the most characteristic changes are the red tongue, swollen lips with vertical cracking and bleeding. The mucosa of the mouth and throat may be bright red, and the tongue may have a typical "strawberry tongue" appearance (marked redness with prominent gustative papillae).

These mouth symptoms are caused by the typical necrotizing microvasculitis with fibrinoid necrosis. Cervical lymphadenopathy is seen in 50% to 75% of people. In the acute phase of the disease, changes in the peripheral extremities can include erythema of the palms and soles, which is often striking with sharp demarcation and often accompanied by painful, brawny edema of the dorsa of the hands or feet. The most common skin manifestation is a diffuse macular-papular erythematous rash, which is quite nonspecific.

5. Diagnosis:

Classically, five days of fever plus four of five diagnostic criteria must be met to establish the diagnosis. The criteria are:

- ✓ erythema of the lips or oral cavity or cracking of the lips
- ✓ rash on the trunk
- ✓ swelling or erythema of the hands or feet
- ✓ red eyes (conjunctival injection)
- ✓ swollen lymph node in the neck of at least 15 mm

Many children, especially infants, eventually diagnosed with Kawasaki disease, do not exhibit all of the above criteria. In fact, many experts now recommend treating for Kawasaki disease even if only three days of fever have passed and at least three diagnostic criteria are present, especially if other tests reveal abnormalities consistent with Kawasaki disease. In addition, the diagnosis can be made purely by the detection of coronary artery aneurysms in the proper clinical setting.

6. Investigations:

A physical examination will demonstrate many of the features listed above.

Blood Tests:

- ✓ Complete blood count may reveal normocytic anemia and eventually thrombocytosis.
- ✓ Erythrocyte sedimentation rate will be elevated.
- ✓ C-reactive protein will be elevated.
- ✓ Liver function tests may show evidence of hepatic inflammation and low serum albumin levels.[104]

Other Optional Tests Include:

- ✓ Electrocardiogram may show evidence of ventricular dysfunction or, occasionally, arrhythmia due to myocarditis.
- ✓ Echocardiogram may show subtle coronary artery changes or, later, true aneurysms.
- ✓ Ultrasound or computerized tomography may show hydrops (enlargement) of the gallbladder.
- ✓ Urinalysis may show white blood cells and protein in the urine (pyuria and proteinuria)
- ✓ without evidence of bacterial growth.
- ✓ Lumbar puncture may show evidence of aseptic meningitis.
- ✓ Angiography was historically used to detect coronary artery aneurysms, and remains the gold standard for their detection.

7. Treatment:

Children with Kawasaki disease should be hospitalized and cared for by a physician who has experience with this disease. When in an academic medical center, care is often shared between pediatric cardiology, pediatric rheumatology, and pediatric infectious disease specialists (although no specific infectious agent has been identified as yet). Treatment should be started as soon as the diagnosis is made to prevent damage to the coronary arteries.

Intravenous immunoglobulin (IVIG) is the standard treatment for Kawasaki disease and is administered in high doses with marked improvement usually noted within 24 hours. If the fever does not respond, an additional dose may have to be considered. In rare cases, a third dose may be given to the child. IVIG by itself is most useful within the first seven days of onset of fever, in terms of preventing coronary artery aneurysm.

Salicylate therapy, particularly aspirin, remains an important part of the treatment (though questioned by some) but salicylates alone are not as effective as IVIG. Aspirin therapy is started at high doses until the fever subsides, and then is continued at a low dose when the patient returns home, usually for two months to prevent blood clots from forming. Except for Kawasaki disease and a few other indications, aspirin is otherwise normally not recommended for children due to its association with Reye's syndrome. Because children with Kawasaki disease will be taking aspirin for up to several months, vaccination against varicella and influenza is required, as these infections are most likely to cause Reye's syndrome. High-dose aspirin is associated with anemia and does not confer benefit to disease outcomes.

8. Prognosis:

With early treatment, rapid recovery from the acute symptoms can be expected, and the risk of coronary artery aneurysms is greatly reduced. Untreated, the acute symptoms of Kawasaki disease are self-limited (*i.e.* the patient will recover eventually), but the risk of coronary artery involvement is much greater. Overall, about 2% of patients die from complications of coronary vasculitis. Patients who have had Kawasaki disease should have an echocardiogram initially every few weeks, and then every one or two years to screen for progression of cardiac involvement.

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