

Kawasaki syndrome

A non-specific disease, with no apparent infectious agent, that affects the mucus membranes, lymph nodes, lining of the blood vessels, and the heart.

General information about Kawasaki syndrome

The cause of Kawasaki disease has not been determined. The incidence is high in Japan where the disease was first described and is recognized now more frequently in the United States. Other risk factors than age are unknown. Kawasaki disease is predominantly a disease of young children, with 80% of patients younger than 5 years of age.

Kawasaki disease is a poorly understood illness. It appears in many respects to be an immune vasculitis (an autoimmune disorder). It is precipitated by unknown outside factors. The disorder affects the mucus membranes, lymph nodes, lining of the blood vessels and the heart. The cardiac involvement and complications are, by far, the most important aspect of the disease. Kawasaki disease can cause vasculitis (inflammation of blood vessels) in the coronary arteries and subsequent coronary artery aneurysms. These aneurysms can lead to myocardial infarction (heart attack) even in young children (rarely). About 20 - 40% of children with Kawasaki disease will have evidence of vasculitis with cardiac involvement.

Kawasaki disease often begins with a high and persistent fever that is not very responsive to normal doses of acetaminophen or ibuprofen. The fever may persist steadily for up to two weeks. The children develop red eyes, red mucous membranes in the mouth, red cracked (fissured) lips, a "strawberry tongue", and swollen lymph nodes. Skin rashes may occur early in the disease and peeling of the skin in the groin (genital area), hands, and feet (especially around the nails and on the palms and soles) may occur.

The changes in the coronary arteries can only be demonstrated by testing. Echocardiography (non-invasive) or angiography, a study in which dye is injected into the blood stream and the heart and its coronary arteries viewed or X-ray may be used.

Causes & symptoms of Kawasaki syndrome

The specific cause of Kawasaki syndrome is unknown, although the disease resembles infectious illnesses in many ways. It has been suggested that Kawasaki syndrome represents an allergic reaction or other unusual response to certain types of infections. Some researchers think that the syndrome may be caused by the interaction of an immune cell, called the T cell, with certain poisons (toxins) secreted by bacteria.

Kawasaki syndrome has an abrupt onset, with fever as high as 104°F (40°C) and a rash that spreads over the patient's chest and genital area. The fever is followed by a characteristic peeling of the skin beginning at the fingertips and toenails. In addition to the body rash, the patient's lips become very red, with the tongue developing a "strawberry" appearance. The palms, soles, and mucous membranes that line the eyelids and cover the exposed portion of the eyeball (conjunctivae) become purplish-red and swollen. The lymph nodes in the patient's neck may also become swollen. These symptoms may last from two weeks to three months, with relapses in some patients.

In addition to the major symptoms, about 30% of patients develop joint pains or arthritis, usually in the large joints of the body. Others develop pneumonia, diarrhea, dry or cracked lips, jaundice, or an inflammation of the membranes covering the brain and spinal cord (meningitis). A few patients develop symptoms of inflammation in the liver (hepatitis), gallbladder, lungs, or tonsils.

About 20% of patients with Kawasaki syndrome develop complications of the cardiovascular system. These complications include inflammation of the heart tissue (myocarditis), disturbances in heartbeat rhythm (arrhythmias), and areas of blood vessel dilation (aneurysms) in the coronary arteries. Other patients may develop inflammation of an artery (arteritis) in their arms or legs. Complications of the heart or arteries begin to develop around the tenth day after the illness begins, when the fever and rash begin to subside. A few patients may develop gangrene, or the death of soft tissue, in their hands and feet. The specific causes of these complications are not yet known.

Diagnosis of Kawasaki syndrome

Because Kawasaki syndrome is primarily a disease of infants and young children, the disease is most likely to be diagnosed by a pediatrician. The physician will first consider the possible involvement of other diseases that cause fever and skin rashes, including scarlet fever, measles, Rocky Mountain spotted fever, toxoplasmosis (a disease carried by cats), juvenile rheumatoid arthritis, and a blistering and inflammation of the skin caused by reactions to certain medications (Stevens-Johnson syndrome).

Once other diseases have been ruled out, the patient's symptoms will be compared with a set of diagnostic criteria. The patient must have a fever lasting five days or longer that does not respond to antibiotics, together with four of the following five symptoms:

Inflammation of the conjunctivae of both eyes with no discharge

**At least one of the following changes in the mucous membranes of the mouth and throat:
"strawberry" tongue; cracked lips; or swollen throat tissues**

At least one of the following changes in the hands or feet: swelling caused by excess fluid in the tissues; peeling of the skin; or abnormal redness of the skin

A skin eruption or rash associated with fever (exanthem) on the patient's trunk

Swelling of the lymph nodes in the neck to a size greater than 1.5 cm.

Since the cause of Kawasaki syndrome is unknown, there are no laboratory tests that can confirm the diagnosis. The following test results, however, are associated with the disease:

Blood tests show a high white blood cell count, high platelet count, a high level of protein in the blood serum, and mild anemia.

Chest x ray may show enlargement of the heart (cardiomegaly).

Urine may show the presence of pus or an abnormally high level of protein.

An electrocardiogram may show changes in the heartbeat rhythm.

In addition to these tests, it is important to take a series of echocardiograms during the course of the illness because 20% of Kawasaki patients will develop coronary aneurysms or arteritis that will not appear during the first examination.

Treatment

Kawasaki syndrome is usually treated with a combination of aspirin, to control the patient's fever and skin inflammation, and high doses of intravenous immune globulin to reduce the possibility of coronary artery complications. Some patients with heart complications may be treated with drugs that reduce blood clotting or may receive corrective surgery.

Follow-up care includes two to three months of monitoring with chest x rays, electrocardiography, and echocardiography. Treatment with aspirin is often continued for several months.

Prognosis

Most patients with Kawasaki syndrome will recover completely, but about 1-2% will die as a result of blood clots forming in the coronary arteries or as a result of a heart attack. Deaths are sudden and unpredictable. Almost 95% of fatalities occur within six months of infection, but some have been reported as long as 10 years afterward. Long-term follow-up of patients with aneurysms indicates that about half show some healing of the aneurysm. The remaining half has a high risk of heart complications in later life.