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# A case of recurrent Kawasaki disease

**Ronald P. Januchowski, DO, Sheron Abraham, OMS IV**

*From the Center for Family Medicine, Spartanburg Regional Medical Center, Spartanburg, SC.*

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Vasculitis;
Mucocutaneous lymph node syndrome

Kawasaki disease is rare form of vasculitis especially affecting the coronary arteries of children under the age of five years. There is no known infectious agent of origination associated with the vasculitis. We report a case of recurrent Kawasaki disease, which makes it a more unusual event because the incidence of recurrent Kawasaki disease in the United States is 0.8%. The patient was a two-year, ten-month-old African-American female with a one-week history of high fever, sore throat, redness of both eyes, nasal congestion, swollen hands and feet, blotchy skin, and desquamation of the genital area. The child was diagnosed with Kawasaki disease and started on treatment that included a dose of intravenous immunoglobulin and high-dose aspirin. Recurrent Kawasaki disease should be considered in children who present with a persistent fever of unknown origin for more than five days and have a previous history of Kawasaki disease. Kawasaki disease can cause dramatic effects on the cardiovascular system if not identified and treated promptly and aggressively. Compared with the initial presentation, recurrent Kawasaki disease has an increased risk of causing coronary artery aneurysms, thrombosis, rupture, myocarditis, pericarditis, and myocardial damage along with sudden death.


Kawasaki disease is classified as a rare form of vasculitis especially affecting the coronary arteries of children under the age of five years with no known infectious agent of origination. The clinical diagnosis of Kawasaki disease is made on the basis of a five-day history of persistent fever of unknown origin that is resolved after empirical treatment with antibiotics, and clinical presentation of at least five of the diagnostic criteria.[1] Kawasaki disease, also known as *mucocutaneous lymph node syndrome*, is a rare diagnosis found most often among children of Japanese descent. It affects 150 per 100,000 children of Japanese descent younger than five years, and in the United States it affects 10 to 15 children per 100,000 younger than five years.[2] This disease, if diagnosis is delayed, is a leading cause of cardiac catastrophe and sudden death in young children via coronary artery aneurysm.[3] Recurrent Kawasaki disease usually occurs on average between two to four years from initial diagnosis. The incidence of recurrent Kawasaki disease worldwide is 2.3%[3] and in the United States 0.8%.[4] In this case report, the patient presented with a recurrence of Kawasaki disease five months after initial diagnosis.

## Case summary

The patient in this case report was a two-year, ten-month-old African-American female that presented to the emergency department with a one-week history of high fever, with a maximum temperature of 103°F; sore throat; bilateral eye conjunctivitis; nasal congestion; swollen hands and feet; blotchy skin; and desquamation of the genital area. The child also had a history of no solid food intake for one week, but adequate fluid intake per the patient's mother. In addition, the child had two to three watery diarrhea episodes daily for three days with a decreased number of wet diapers. The patient's mother stated symptoms were identical to

Corresponding author: Ronald P. Januchowski, DO, VCOM, Family Medicine Program, Spartanburg Regional Medical Center, 853 N. Church Street, Suite 510, Spartanburg, SC 29303-3077.
E-mail address: rjanuchowski@srhs.com.

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when the patient was first diagnosed with Kawasaki disease, five months before this presentation. The patient was treated with intravenous gamma globulin and oral aspirin and recovered without sequelae at that time.

The initial physical examination revealed an ill-appearing febrile child. The patient weighed 12.7 kilograms. Her vital signs included an oral temperature of 101.5°F, a heart rate of 155 beats per minute, a respiratory rate of 18 breaths per minute, and a blood pressure of 105/63 mm Hg. The patient had bilateral nonexudative conjunctivitis; erythematous lips and oral mucosa; and dry, cracked lips. Slight edema and erythema of the hands and feet were noted bilaterally. No prominent cervical lymph nodes were palpated. There was desquamation of the genital area. There were no abnormalities noted on cardiovascular, pulmonary, or gastrointestinal physical examination. Complete laboratory data is found in Fig. 1.

Prophylactic antibiotics were initiated to cover possible meningitis. These antibiotics were discontinued once lumbar puncture results were negative for bacterial meningitis.

Given the laboratory studies, radiographs, and echocardiogram results, the differential diagnosis was revised to include fever of unknown origin and aseptic meningitis, with recurrent Kawasaki disease as the most probable diagnosis.

The patient was treated with the standard medication regimen for Kawasaki disease (see discussion below) and subsequently had clinical improvement with treatment.

The patient was discharged three days after admission. Physical examination on the day of discharge was normal with the exception of mild, dry lips and mild desquamation of the genital area. The patient received a six-week prescription for aspirin therapy and her parent was asked to follow up with their primary care physician in one week and a pediatric cardiologist for a repeat echocardiogram several weeks later.

## Discussion

As this case illustrates, recurrent Kawasaki disease was considered in this young child who presented with persistent

- Complete Blood Count: White blood cells 29,100 cells/mm$^3$ differential of 85% segmented neutrophils, 1% bands, 8% lymphocytes, 5% monocytes, 1% eosinophils. Platelets 518,000 per mm$^3$.
- Partial Thromboplastin Time and Prothrombin Time normal.
- C -Reactive Protein 18.7.
- Urinalysis positive for protein and moderate trace of blood. Urine culture negative.
- Blood culture negative. Throat culture, Rapid Strep, Antistreptolysin O and Rapid Influenza all negative.
- Lumbar Puncture: Negative organism growth in 48 hrs.
- Chest X-ray evident for under expanded lungs with hazy basilar opacity suggestive of atelectasis.
- Echocardiography: Heart size and function normal; no coronary artery dilatation. Trace of mitral regurgitation, aortic regurgitation, tricuspid valve regurgitation, pulmonic valve regurgitation; no pleural effusion; estimated right ventricular pressure 22mmHg; Normal right and left coronary artery origins; left main coronary artery 1.9 mm and right coronary artery 1.3-2.0 mm.

**Figure 1** Patient laboratory and imaging data.

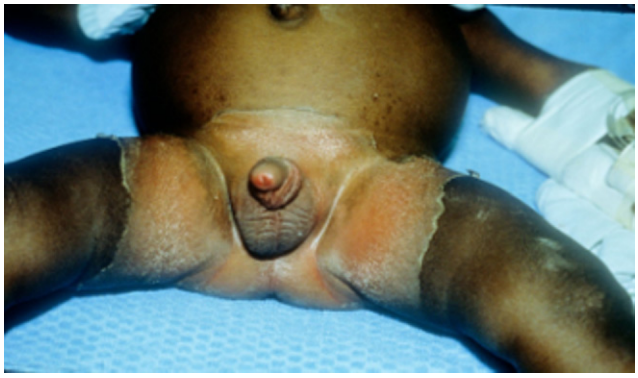

**Figure 2** Desquamation of the genitalia. (Reprinted with the permission from Dr. Jim Thomas at dermnet.com.)

fever (>102°F) of unknown origin for more than five days and had been diagnosed previously with Kawasaki disease in the last 12 months.[5]

This disease can cause dramatic effects on the cardiovascular system if not identified and treated promptly and aggressively. Recurrent Kawasaki disease has an increased risk of causing coronary artery aneurysms, thrombosis, rupture, myocarditis, pericarditis, and myocardial damage, along with sudden death compared with the first presentation of the disease.[5]

Kawasaki disease is a clinical diagnosis based on presentation of persistent fever for at least five days and four of five of the following characteristics:

1. Polymorphous rash
2. Characteristic changes in extremities to include erythema or desquamation of genital area, fingers, or toes (Fig. 2)
3. Bilateral conjunctivitis
4. Cervical lymphadenopathy
5. Oropharyngeal changes including "strawberry" tongue or dry, erythematous, cracked lips (Fig. 3).[5,6]

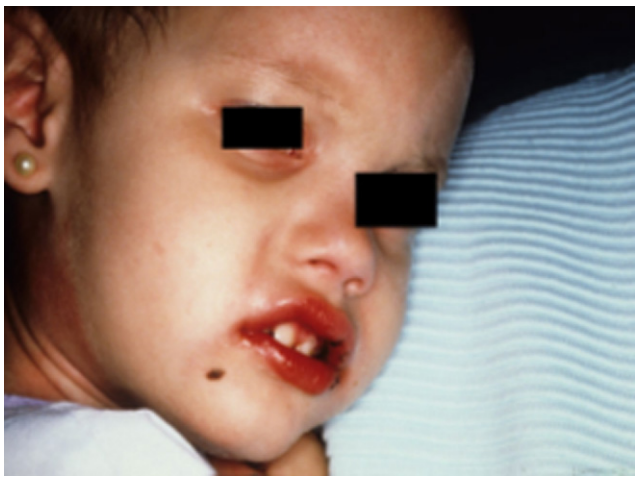

**Figure 3** Erythema and edema of the lips. (Reprinted with the permission from Dr. Jim Thomas at dermnet.com.)

This patient's clinical presentation with a persistent fever, edematous hands and feet, erythematous lips and oral mucosa, and desquamation of genitals supported the diagnosis of Kawasaki disease. Because of the patient's history of a previous diagnosis within the previous 12 months, the diagnosis of recurrent Kawasaki disease was made.

The diagnosis of Kawasaki disease has no definitive radiologic or laboratory test. The differential diagnosis would include aseptic or bacterial meningitis, scarlet fever, juvenile rheumatoid arthritis, drug reactions, and other infectious processes. Physicians should exclude these other disease processes as needed with a complete blood count, blood cultures, urine cultures, chest radiograph, or lumbar puncture based on clinical symptoms. Typical laboratory findings in Kawasaki disease include elevation of acute-phase reactants (ie, white blood count with left shift, erythrocyte sedimentation rate, C-reactive protein) with negative cultures. In this case, the clinical manifestations, nonspecific laboratory findings, and previous diagnosis of Kawasaki disease all support suspicion for recurrent Kawasaki disease.

Once a diagnosis has been made, the patient should be treated using the standard medication regimen for Kawasaki disease that includes intravenous gamma globulin and oral aspirin. Standard treatment includes a single dose of intravenous immunoglobulin (IVIG), 2 g/kg body weight over 12 hours, which has shown to reduce the incidence and severity of aneurysm formation and symptomatic relief for the acute illness.[1] Although IVIG is not entirely without risk, its benefits outweigh the risks in patients with confirmed Kawasaki disease. Aspirin is initially administered at high-dose intervals for two days at 80–100 mg/kg body weight per day divided into four intervals. This dose is continued until the patient has been afebrile for 48 hours, and then the dose is decreased to 3–5 mg/kg per day for six to eight weeks for its antiinflammatory and antithrombotic effects.[1] Aspirin may be continued indefinitely in patients who develop coronary abnormalities as a result of Kawasaki disease. Physicians need to inform the caregivers for these patients about the possible risk of Reyes syndrome if aspirin therapy is continued.

Current research states that baseline echocardiography and an electrocardiogram should be obtained one to two weeks after presentation of the disease and four to six weeks after the treatment phase to confirm that there is no damage to cardiac vasculature and cardiac muscle. Most anatomical structural damage tends to occur one to two weeks after symptom onset. The ability to recognize the signs and symptoms of anatomical structure damage, such as the development of coronary artery aneurysms, is critical to prevent further morbidity of the disease process. Depending on the level of structural damage to the coronary arteries, it may be necessary to place the patient on physical activity restriction. At the minimum, the patient should undergo cardiovascular risk assessment and counseling at five-year intervals. This follow-up schedule would obviously need to be modified if the patient is in a higher risk group, and may include invasive cardiac testing. A pediatric cardiologist should be consulted to assess risk and provide further testing options as needed.

In summary, it is imperative to recognize the signs and symptoms of Kawasaki disease and be able to make the clinical diagnosis in a timely manner. Making this diagnosis and providing treatment reduces the long term sequelae associated with the disease. It is also important to have an understanding of the effects of Kawasaki disease on the cardiac muscle and vasculature in clinical practice. The rare presentation of recurrent Kawasaki disease should remain in the differential diagnosis of a pediatric patient presenting with fevers, rash, and a previous history of Kawasaki disease.

## References

1. Newburger JW, Takahashi M, Gerber MA, et al: Diagnosis, treatment, and long-term management of Kawasaki disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. Circulation 110:2747-2771, 2004
2. Freeman AF, Shulman ST: Kawasaki disease: summary of the American Heart Association guidelines. Am Fam Physician 74:1141-1148, 2006
3. Hsieh YC, Wu MH, Wang JK, et al: Clinical features of atypical Kawasaki disease. J Microbiol Immuno Infect 35:57-60, 2002
4. Gupta-Malhotra M, Rao PS: Current perspectives on Kawasaki's disease. Indian J Pediatr 72:621-629, 2005
5. Pemberton MN, Doughty IM, Middlehurst RM, et al: Recurrent Kawasaki disease. Br Dent J 186:270-271, 1999
6. Newburger JW, Takahashi M, Gerber MA, et al: Diagnosis, treatment and long-term management of Kawasaki disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. Pediatrics 114:1708-1733, 2004