Case In Point
An Intriguing Diagnosis

Atypical Kawasaki Disease in an 11-Year-Old Boy

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An 11-year-old Caucasian boy with intermittent fever for the past 5 days presented to the local school-based health center accompanied by his mother. The mother reported that her son had spikes in temperature up to 40.3°C (104.5°F), without associated symptoms. He was initially evaluated by a local physician who diagnosed a “virus” and was sent home with instructions to use antipyretics as needed. The fever persisted and by the morning of presentation a nonpruritic rash had developed on the lower extremities. No family members in the household had similar symptoms. The child took no medications other than acetaminophen and ibuprofen and had no drug or environmental allergies. Past medical history was unremarkable.

Physical examination revealed an alert, healthy-appearing boy. Vital signs, including temperature (36.8°C [98.3°F]) and heart rate (76 beats per minute), were normal. His lips appeared full with a prominent red-purple hue, and multiple red petechial lesions were noted along the hard palate and buccal mucosa (Figure 1). Cervical lymphadenopathy was not evident. A pink-red, papular eruption was noted along the lower abdomen, proximal thighs, and buttocks (Figure 2). Some swelling of the hands and feet was noted. Abdomen was soft and non-

Figure 1 – The patient had full lips with a prominent red-purple hue (A) and multiple red petechial lesions along the hard palate and buccal mucosa (B). Oral mucosal changes in classic Kawasaki disease may include strawberry tongue; dry cracked lips; and erythema of lips, mouth, and pharynx.
tender. Neurological examination revealed no abnormalities.

The patient was immediately referred to a nearby children's hospital for further workup for suspected atypical Kawasaki disease. During the initial days of hospitalization, the patient became febrile, his skin eruption became generalized, and he was treated for conjunctivitis. Laboratory studies showed the following: white blood cell count, 16,000/µL; C-reactive protein, 6.84 mg/L; erythrocyte sedimentation rate, 121 mm/h; albumin, 2.6 g/dL; and alanine aminotransferase, 112 U/L. Other test results were normal.

An echocardiogram showed no significant abnormalities. The pediatric cardiology consultant agreed with the diagnosis of atypical Kawasaki disease.

Intravenous immunoglobulin (IVIG) and high-dose aspirin therapy were initiated (IVIG and high-dose aspirin therapy 80 mg/kg/d divided in 4 doses) were initiated. The fever and symptoms subsequently resolved, and the patient was discharged home after an 8-day hospitalization.

KAWASAKI DISEASE: AN OVERVIEW

Kawasaki disease is an acute febrile vasculitis first described by the Japanese physician Tomisaku Kawasaki in 1967. The disease occurs worldwide, with the highest incidence in Asians and the lowest incidence in Caucasians. Approximately 80% of cases occur in those younger than 5 years. Rarely the disease may occur in adolescents and adults. About 3000 cases are diagnosed in the United States annually. It is the leading cause of acquired heart disease among children in developed countries.

The complications of untreated Kawasaki disease can be fatal. It is imperative that the diagnosis be established promptly so that treatment with IVIG and high-dose aspirin therapy may be provided. Coronary artery sequelae—usually aneurysms—with the potential for thrombosis, stenosis, and sudden death may occur in 20% of untreated cases of Kawasaki disease. An echocardiogram is required to identify coronary artery morphology and to assess cardiac function and coronary artery dimensions.

FINDINGS IN ATYPICAL KAWASAKI DISEASE

Patients with atypical or incomplete Kawasaki disease lack all of the diagnostic criteria of classic Kawasaki disease. For a diagnosis of atypical Kawasaki disease, the patient must have:

- Fever for 5 or more days.
- Two of 5 clinical features in classic cases (including conjunctival injection, oral mucosal changes, erythema and swelling of hands and feet, erythematous rash on trunk and extremities, and cervical adenopathy).
- C-reactive protein greater than 3.0 mg/L and/or erythrocyte sedimentation rate greater than 40 mm/h.
- Compatible laboratory findings (at least 3 of the following: albumin, ≥ 3.0 g/dL; anemia for age; elevation of alanine aminotransferase; platelets after seventh day of illness, ≥ 450,000/µL; white blood cell count, ≥ 15,000/µL; urine, ≥ 10 white blood cells/high-powered field) or positive echocardiogram.

TAKE-HOME MESSAGE

The clinician should consider Kawasaki disease in the differential diagnosis when the clinical signs and symptoms of the disease are evident in patients older than 5 years.

REFERENCES:


Figure 2 – The patient had a pink-red, papular eruption along the lower abdomen, proximal thighs, and buttocks. The rash of classic Kawasaki disease is marked by erythema and may appear maculopapular, scarlatiniform, or erythema multiforme–like. Peeling in the groin may be present.