With Kawasaki Disease, Time is Coronary Health

BY CLAIRE JANSSON-KNODELL AND RHAMY MAGID, M.D.

A previously healthy 3-year-old Hmong boy presented to Children’s Hospitals and Clinics of Minnesota with a history of fever that was unremitting despite antipyretics. Two weeks prior to admission at Children’s, he presented to an outside hospital with a fever accompanied by left-sided neck swelling. A neck ultrasound showed a lymph node measuring 3.5 x 2.7 x 2.2 cm, without abscess or fluid collection. He was treated for acute cervical lymphadenitis with antibiotics (ceftriaxone initially, then ampicillin-sulbactam). His undulating fevers continued, but he was discharged after two days with oral antibiotics (amoxicillin-clavulanate). At home, his family discontinued this medication after his mother noticed a spotted rash on his shins believed to be an allergic reaction. During a follow-up visit to his pediatrician, the boy had a low-grade fever, swelling in his legs, and an erythematous rash. He was referred to the hospital, but his mother chose to keep him home because she assumed he had improved. He was “playful and interactive” at home and the neck swelling had lessened. A week later, the child returned to the clinic with persistent fever and pain in his foot that caused him to limp. The child also had red eyes, which his mother attributed to frequent crying. The pediatrician sent the boy directly to Children’s Hospital for admission.

On exam, the boy had a fever of 103.1°F and was tachycardic (161 beats per minute). His blood pressure was 110/66 mm Hg, and he was irritable. He weighed 15.2 kg. His sclerae were injected bilaterally without exudate. His lips appeared bright pink with cheilosis without frank cracking. His oropharynx was erythematous. He did not have cervical lymphadenopathy. His hands and feet were edematous bilaterally. He had no rash, petechiae or ecchymosis. He had nonbleeding desquamation of his hands circumferentially around his thumbs.

The boy had a WBC of 25,000/mm³ with a neutrophil predominance, hemoglobin level of 8.1g/dL and a platelet count of 895,000/mm³. He had a mild transaminitis with an alanine aminotransferase level of 60 U/L. His C-reactive protein was 9.48 mg/dL and his erythrocyte sedimentation rate was >140 mm/hour.

This boy’s history, physical exam and lab values were suggestive of Kawasaki disease (Table).

Because of his late presentation, there was concern for coronary aneurysm. He was immediately started on intravenous immune globulin therapy (IVIG) and high-dose aspirin.

Transthoracic echocardiogram showed three aneurysms—one in each of his coronary arteries. The aneurysm in the left circumflex artery measured 5.6 mm, a saccular aneurysm in the right coronary artery was 6.5 mm in size, and the giant

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**Clinical Criteria for Diagnosis of Kawasaki Disease**

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<th>Fever of at least five days</th>
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<td>Presence of four or more of the following clinical features:</td>
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<td>* Conjunctival injection: bilateral, painless, without exudate</td>
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<td>* Cervical lymphadenopathy: &gt;1.5 cm, usually unilateral</td>
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<td>* Oral mucosal changes: erythema and cracking of lips, strawberry tongue, diffuse injection of oral and pharyngeal mucosa</td>
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<td>* Polymorphous rash</td>
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<td>* Changes in extremities: acute stage — erythema and edema of hands and feet, convalescent stage — membranous desquamation of fingertips</td>
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Exclusion of alternative diagnosis

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Learning points

- Early recognition is key to preventing cardiovascular sequelae.
- Treatment within the first 10 days of illness is critical to the prevention of coronary aneurysms, reducing the risk fivefold.
- If there is concern regarding patient compliance in cases where the index of suspicion for Kawasaki disease is high, the seriousness should be clearly communicated to the family and close follow up may be beneficial.

Discussion

This case illustrates the potential for severe consequences when there is late diagnosis of Kawasaki disease. Without IVIG treatment, about 25% of children with Kawasaki disease develop an aneurysm; with IVIG administration that number is reduced to 4%. Recognition of this constellation of findings as Kawasaki disease is crucial to the administration of appropriate IVIG therapy for prevention of aneurysms in coronary arteries. Additionally, this case highlights the value of culturally appropriate care. Perhaps if the gravity of the situation was communicated to the family in a way that was clearly understood, this poor outcome could have been avoided. MM

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REFERENCES