CASE REPORT

THE PATIENT
3-year-old girl

SIGNS & SYMPTOMS
– Fever
– Cervical lymphadenopathy
– Leukocytosis

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THE CASE

A previously healthy 3-year-old girl presented to the emergency department with 4 days of fever and 2 days of right-side neck pain. The maximum temperature at home was 103 °F. The patient was irritable and vomited once. There were no other apparent or reported symptoms.

The neck exam was notable for nonfluctuant, swollen, and tender lymph nodes on the right side. Her sclera and conjunctiva were clear, and her oropharynx was unremarkable. Lab work revealed leukocytosis, with a white blood cell (WBC) count of 15.5 × 10^3/μL (normal range, 4.0-10.0 × 10^3/μL). She was given one 20 cc/kg normal saline bolus, started on intravenous clindamycin for presumed cervical lymphadenitis, and admitted to the hospital.

On Day 2, the patient developed a fine maculopapular rash on her chest, abdomen, and back. She had spiking fevers—as high as 102.2 °F—despite being on antibiotics for more than 24 hours. The erythrocyte sedimentation rate (ESR) was 39 mm/h (0-20 mm/h), and C-reactive protein (CRP) was 71.4 mg/L (0.0-4.9 mg/L). Due to concern for abscess, a neck ultrasound was performed; it showed a chain of enlarged lymph nodes in the right neck (largest, 2.3 × 1.1 × 1.4 cm) and no abscess.

On Day 3, clindamycin was switched to intravenous ampicillin/sulbactam because a nasal swab for methicillin-resistant *Staphylococcus aureus* was negative. A swab for respiratory viral infections was also negative. The patient then developed notable facial swelling, bilateral bulbar conjunctival injection with limbic sparing, and swelling of her hands and feet.

THE DIAGNOSIS

By the end of Day 3, the patient's lab studies demonstrated microcytic anemia and low albumin (2.5 g/dL), but no transaminitis, thrombocytosis, or sterile pyuria. An electrocardiogram was unremarkable. A pediatric echocardiogram revealed hyperemic coronaries, indicating inflammation. The coronary arteries were measured in the upper limits of normal, and the patient's Z-scores were < 2.5. (A Z-score < 2 indicates no involvement, 2 to < 2.5 indicates dilation, and ≥ 2.5 indicates aneurysm abnormality.[1]) An ultrasound of the right upper quadrant revealed an enlarged/elongated gallbladder. The patient therefore met clinical criteria for Kawasaki disease.

DISCUSSION

Kawasaki disease is a self-limited vasculitis of childhood and the leading cause of acquired heart disease in children in developed countries.[1] The annual incidence of Kawasaki disease in North America is about 25 cases per 100,000 children < 5 years of age.[1] In the United States, incidence is highest in Asian and Pacific Islander populations (30 per 100,000) and is particularly high among those of Japanese ancestry (~200 per 100,000).[2] Disease prevalence

is also noteworthy in Non-Hispanic African American (17 per 100,000) and Hispanic (16 per 100,000) populations.[2]

Diagnosis of Kawasaki disease requires presence of fever lasting at least 5 days and at least 4 of the following: bilateral bulbar conjunctival injection, oral mucous membrane changes (erythematous or cracked lips, erythematous pharynx, strawberry tongue), peripheral extremity changes (erythema of palms or soles, edema of hands or feet, and/or periungual desquamation), diffuse maculopapular rash, and cervical lymphadenopathy (≥ 1.5 cm, often unilateral). If ≥ 4 criteria are met, Kawasaki disease can be diagnosed on the fourth day of illness.[1]

Laboratory findings suggesting Kawasaki disease include a WBC count ≥ 15,000/mcL, normocytic, normochromic anemia, platelets ≥ 450,000/mcL after 7 days of illness, sterile pyuria (≥ 10 WBCs/high-power field), serum alanine aminotransferase level > 50 U/L, and serum albumin ≤ 3 g/dL.

Cardiac abnormalities are not included in the diagnostic criteria for Kawasaki disease but provide evidence in cases of incomplete Kawasaki disease if ≥ 4 criteria are not met and there is strong clinical suspicion.[1] Incomplete Kawasaki disease should be considered in a patient with a CRP level ≥ 3 mg/dL and/or ESR ≥ 40 mm/h, ≥ 3 supplemental laboratory criteria, or a positive echocardiogram.[1]

Ultrasound imaging may reveal cervical lymph nodes resembling a "cluster of grapes."[3] The case patient's imaging showed a "chain of enlarged lymph nodes." She likely had gallbladder "hydrops" due to its increased longitudinal and horizontal diameter and lack of other anatomic changes.[4]

Prompt treatment is essential

Treatment for complete and incomplete Kawasaki disease is a single high dose of intravenous immunoglobulin (IVIG) along with aspirin. Patients meeting criteria should be treated as soon as the diagnosis is established.[5] A single high dose of IVIG (2 g/kg), administered over 10 to 12 hours, should be initiated within 5 to 10 days of disease onset. Administering IVIG in the acute phase of Kawasaki disease reduces the prevalence of coronary artery abnormalities.[6] Corticosteroids may be used as adjunctive therapy for patients with high risk of IVIG resistance.[1,7-9]

Our patient was not deemed to be at high risk for IVIG resistance (Non-Japanese patient, age at fever onset > 6 months, absence of coronary artery aneurysm[9]) and was administered IVIG on Day 4. She was also given moderate-dose aspirin, then later transitioned to low-dose aspirin. The patient's fevers improved, she was less irritable, and she had periods of playfulness. Physical exam then showed erythematous and cracked lips with peeling skin.

The patient was discharged home on Day 8, after her fever resolved, with instructions to continue low-dose aspirin and to obtain a repeat echocardiogram, gallbladder ultrasound, and lab work in 2 weeks. At her follow-up appointment, periungual desquamation was noted, and ultrasound showed continued enlarged/elongated gallbladder. A repeat echocardiogram was not available. Overall, the patient recovered from Kawasaki disease after therapeutic intervention.

THE TAKEAWAY

Kawasaki disease can be relatively rare in North American populations, but it is important for physicians to be able to recognize and treat it. Untreated children have a 25% chance of developing coronary artery aneurysms.[1,10,11] Early treatment with IVIG can decrease risk to 5%, resulting in an excellent medium- to long-term prognosis for patients.[10] Thorough physical examination and an appropriate degree of clinical suspicion was key in this case of Kawasaki disease. JFP

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Untreated children with Kawasaki disease have a 25% chance of developing coronary artery aneurysms.

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