A 7-Year-Old Boy with Abdominal Pain, Fever, and Rash

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A 7-year-old boy presented to our emergency department with 5 days of abdominal pain and 3 days of fever, rash, and emesis. He had no jaundice, diarrhea, or constipation.

His abdominal examination was significant for an exquisitely tender right upper-quadrant but without signs of peritoneal inflammation. He had a scarlatiniform rash on his face, trunk, and extremities, but no Pastia lines. Ocular and oropharyngeal examination were unremarkable, and neither extremity changes nor cervical adenopathy were present.

His laboratory tests were remarkable for a sodium of 132 mmol/L (normal range, 138 mmol/L to 145 mmol/L), alanine aminotransferase of 217 U/L (normal range, 10 U/L to 35 U/L), aspartate aminotransferase of 216 U/L (normal range, 15 U/L to 40 U/L), conjugated bilirubin of 1.1 mg/dL (normal range, 0.0 mg/dL to 0.3 mg/dL), alkaline phosphatase of 439 U/L (normal range, 175 U/L to 420 U/L), gamma-glutamyl transpeptidase of 230 U/L (normal range, 11 U/L to 221 U/L), albumin of 2.6 g/dL (normal range, 3.7 g/dL to 5.6 g/dL), and a C-reactive protein (CRP) of 9 mg/dL (normal range, 0 mg/dL to 0.9 mg/dL).

An infectious workup yielded negative results for human herpesvirus-6, Epstein-Barr virus, adenovirus, parvovirus B19, human metapneumovirus, rhinovirus, influenza A/B, parainfluenza 1/2/3, enterovirus, parechovirus, and *Mycoplasma pneumoniae*. Blood, urine, and gastrointestinal cultures were negative; however, a throat culture ultimately grew *group A streptococcus* (GAS), and thus amoxicillin was started on hospital day 2 for suspected scarlet fever.

An abdominal ultrasound was obtained (see Figure 1A, page 276) and showed a markedly distended gallbladder with sludge but with normal wall thickness and no pericholecystic fluid, calculi, or intra- or extrahepatic biliary duct dilation. This imaging study revealed the cause of his right upper-quadrant pain and helped elucidate his underlying diagnosis.

For diagnosis, see page 276

Editor’s note: Each month, this department features a discussion of an unusual diagnosis in genetics, radiology, or dermatology. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via email at Pediatrics@Healio.com.
**Case Challenge**

**Diagnosis:**

**Gallbladder Hydrops Associated with Kawasaki Disease**

The abdominal ultrasound revealed a markedly distended gallbladder, consistent with gallbladder hydrops (see Figure 1A). Repeat lab tests 48 hours after admission revealed an increase in his peripheral white blood cell count to 16,000 cells/mcL with 70% neutrophils, 11% eosinophils, and 8% bands, as well as an increase in CRP to 16.3 mg/dL. On hospital day 4 he developed bilateral scleral injection consistent with episcleritis. His persistent, severe right upper-quadrant abdominal pain prompted further diagnostic imaging, so an abdominal computed tomography (CT) scan was obtained on hospital day 4 (see Figure 1B) that showed a massively distended gallbladder consistent with gallbladder hydrops, but with no other abdominal pathology. He continued to have daily fevers even 3 days after starting amoxicillin.

Given his prolonged fever (7 days), diffuse rash, gallbladder hydrops, elevated inflammatory markers, and supporting laboratory data (ie, leukocytosis with neutrophilia, eosinophilia and immature forms, anemia, hyponatremia, hypoalbuminemia, and elevated transaminases), intravenous immunoglobulin (IVIG) and high-dose aspirin were initiated on hospital day 5 for a diagnosis of incomplete Kawasaki disease. His right upper-quadrant abdominal pain markedly improved, but he continued to be febrile. A second course of IVIG was administered on day 12 of fever and he defervesced within 24 hours. Serial echocardiograms have shown no signs of coronary artery sequelae.

**DISCUSSION**

Gallbladder hydrops, or acute acalculous distention of the gallbladder, has been associated with many infectious, rheumatologic, and systemic etiologies in the pediatric population. However, pediatric gallbladder hydrops is associated most commonly with Kawasaki disease, with recent estimates of its presence in up to 15% of children with this diagnosis. It typically occurs in the first 2 weeks of illness and can last approximately 2 weeks, but may persist, and most frequently arises in older children. Diagnosis is confirmed with abdominal ultrasound, typically showing a massively distended anechoic, elliptical-shaped gallbladder. Management is supportive and nonoperative unless there are signs of complications, such as rupture, cholecystitis, or peritonitis.

In this case, the finding of gallbladder hydrops aided in making a diagnosis of incomplete Kawasaki disease. Kawasaki disease is a pediatric systemic vasculitis that most typically presents with prolonged fever (≥ 5 days) and at least four of five major supporting signs: 1) polymorphous rash; 2) cervical lymphadenopathy; 3) oral mucous membrane changes; 4) bilateral non-exudative conjunctival injection; or 5) findings of erythema, edema, or desquamation on the extremities. It is the most common cause of pediatric-acquired heart disease in the developed world, and at least 20% of children with Kawasaki disease will develop coronary artery aneurysms if untreated, with cardiovascular consequences ranging from chest pain or arrhythmia to sudden death. However, treatment with IVIG within the first 10 days of illness can prevent coronary artery sequelae.

**Figure 1.** Abdominal ultrasound (A) and computed tomography scan (B) demonstrating a markedly distended, elliptical gallbladder with no other signs of liver or biliary pathology, consistent with gallbladder hydrops.
days of illness significantly reduces the risk of cardiovascular sequelae.5

The diagnosis of Kawasaki disease is often challenging and frequently delayed due to the lack of a specific diagnostic test, similarities with other pediatric diagnoses, and dependence upon a constellation of clinical criteria that may not occur simultaneously.9,10 Furthermore, children with incomplete Kawasaki disease, defined as prolonged fever with only two or three major criteria along with supplementary laboratory and imaging data, pose an additional challenge in diagnosis because few pediatricians are familiar with the supplementary criteria, and yet these patients are also at risk for developing coronary artery sequelae.1,11

Consequently, it is imperative that pediatricians be skilled in recognizing both the major and minor clinical signs of Kawasaki disease in order to accurately identify those children who require further diagnostic workup and close clinical follow-up. This case report highlights one such clinical scenario, in which the presence of gallbladder hydrops aided in making a diagnosis of incomplete Kawasaki disease and initiating timely treatment with IVIG.

The presence of gallbladder hydrops in this case also demonstrates the clinical overlap of GAS infections and Kawasaki disease. Although it has been reported to occur with both disease processes, gallbladder hydrops is much more common in Kawasaki disease.1,3,4,12-14 We suspect that in this case, isolation of GAS from the patient’s throat culture represented asymptomatic colonization, given his lack of exudative pharyngitis, Pastia lines, and response to amoxicillin. Early cases of scarlet fever-associated gallbladder hydrops were reported before Kawasaki disease was recognized in the United States, and in retrospect these cases may have met the clinical criteria for Kawasaki disease.12-14 Not surprisingly, these children continued to have fever and systemic illness on appropriate anti-streptococcal antibiotics.12-14 It is important for pediatricians to consider the diagnosis of Kawasaki disease when fever and inflammation continue despite appropriate anti-streptococcal antibiotics in children diagnosed with GAS infection.

CONCLUSION

Pediatricians should be aware that Kawasaki disease can initially present with gastrointestinal symptoms, such as right upper-quadrant pain from gallbladder hydrops, and should continue to monitor these children for the development of other signs and symptoms consistent with this important pediatric vasculitis. Recognition of this association will aid in prompt diagnosis and treatment of Kawasaki disease and in preventing the long-term cardiac sequelae associated with this common pediatric syndrome.

REFERENCES