Acute acalculous cholecystitis (AAC) is a rare, potentially serious disease that has been associated with Kawasaki disease (KD) in children. Studies suggest that patients presenting with severe abdominal symptoms secondary to KD have increased resistance to intravenous immunoglobulin (IVIG), and a higher rate of coronary artery aneurysms. We describe an eight-year-old boy who presented to the emergency department with severe abdominal pain and was diagnosed with AAC and KD. He was treated with IVIG and high-dose aspirin, achieving good response with complete symptom resolution. He had no coronary artery aneurysms or further complications and was discharged after three days. [Clin Pract Cases Emerg Med. 2019;3(4):383–386.]
(35-104 U/L), and gamma-glutamyl transferase levels of 102 U/L (9-48 U/L). Formal abdominal ultrasonography revealed a distended gallbladder with scant pericholecystic fluid and sludge (Image 1). Computed tomography showed a distended gallbladder (Image 2).

A diagnosis of KD with AAC was made, and the patient was started on oral high-dose aspirin. He was transferred to a tertiary care center where he also received IVIG. All symptoms improved with treatment. His echocardiogram did not show any evidence of coronary artery aneurysms. The patient was discharged home with no further complications after hospital day three.

DISCUSSION
KD is a common cause of pediatric vasculitis. Nonetheless, distinguishing KD from other febrile illness remains one of the biggest challenges that emergency physicians face. In the United States KD has surpassed rheumatic fever as the leading cause of acquired heart disease in children, affecting 19 per 100,000 children under the age of five.

The clinical criteria for the diagnosis of KD include at least five days of fever plus at least four of five principal clinical features: polymorphous rash; oral changes; bilateral conjunctival injection; cervical lymphadenopathy; and extremity changes. If four or five of the clinical criteria are met, physicians may proceed to treatment; however, if only two or three of the five principal clinical criteria are met but clinical suspicion remains high, supplemental laboratory findings may aid diagnosis. Typically, complete blood count, hepatic panel, C-reactive protein level, erythrocyte sedimentation rate, and urinalysis are sufficient to supplement the two or three principal clinical criteria to enable the diagnosis. Such cases are much less common and are often referred to as “incomplete” or “atypical” KD.

Cardiac complications such as coronary artery aneurysm are the main concern with KD; nonetheless, other organ systems can be affected. For example, gastrointestinal (GI) symptoms with hepatobiliary abnormalities are the initial presentation in some patients. Atypical signs and symptoms that should prompt clinical suspicion for KD are hepatic dysfunction, gallbladder hydrops, jaundice, cholestasis, paralytic ileus, and AAC. Yi et al. noted that gallbladder distention alone in patients with KD is associated with coronary artery complications. A 2018 multicenter study conducted in Italy showed that GI symptoms as a manifestation of KD indicate greater risk for severe coronary lesions. Other factors included delayed treatment, low albumin level, and age younger than six months.

Although AAC is uncommon in pediatric patients, recognizing it early is vital given the high incidence of
coronary artery aneurysm associated with it. For a diagnosis of AAC, two of four ultrasonic criteria must be met: gallbladder distention; increased wall thickness (>3.5 mms); presence of sludge; or presence of pericholecystic fluid. This diagnosis can be made by point-of-care ultrasound in the ED.

Some have suggested that medical treatment, rather than immediate surgical intervention, is the preferred way to manage AAC in children with KD. The standard medical treatment for KD is high-dose aspirin (80–100 mg/kg/day) in conjunction with four doses of IVIG. Although aspirin is thought to have antiplatelet and anti-inflammatory effects, it does not reduce the risk for coronary artery aneurysm formation. In contrast, IVIG has been shown to reduce this risk, although it is most effective when administered within 7-10 days of illness onset. Even so, a retrospective study by Chen et al. found that KD patients with AAC treated with IVIG were more likely to be IVIG-resistant than were KD patients without AAC, thus bringing into question whether patients with AAC should receive IVIG therapy.

CONCLUSION

Because KD may present similarly to other benign or potentially deadly diseases, it remains a challenging disease for emergency physicians to recognize. It is imperative that they be aware of the unusual presentations of the disease, such as AAC, and its association with coronary aneurysms. KD must always be considered in the differential diagnosis of a child with prolonged fever. The treatment of AAC in a child with KD is initially medical rather than surgical.

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Documented patient informed consent and/or Institutional Review Board approval has been obtained and filed for publication of this case report.

Image 2. Computed Tomography showing distended gallbladder (arrow).

REFERENCES


