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An Unusual Presentation of Kawasaki Disease

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Abstract We present a patient suffering from Kawasaki disease (KD) with unusual manifestations. Severe myocarditis with shock and gallbladder hydrops dominated the clinical picture in the early phase, when the cardinal features of KD had not yet fully manifested themselves. KD should be included in the differential diagnosis of a febrile child with multi-organ dysfunction.

Key words Gallbladder hydrops; Kawasaki disease; Myocarditis

Introduction

Kawasaki disease (KD) is a febrile condition affecting children. It is noted for its association with vasculitis of coronary vessels. First described by Tomisaku Kawasaki in 1967, more than 140,000 cases have been reported in Japan alone since then.[1,2] The diagnostic criteria include persistent high fever (more than five days by definition), along with four out of five of the following: bilateral non-purulent conjunctivitis, characteristic changes oropharyngeal mucosa (notably injected pharynx, fissured lips and strawberry tongue), polymorphous non-vesicular rash, changes of the extremities (in particular erythema and desquamation) and cervical lymphadenopathy. Various cardiac and non-cardiac associated features or complications were reported over the years (Table 1).[2,3] Here we report a case of KD with unusual presentations.

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Case Report

A 3-year-old Chinese girl presented to the paediatric intensive care unit (PICU) of a tertiary referral centre with progressive jaundice, altered conscious state and shock. She had a five-day history of high fever (up to 40°C)

Table 1 Cardiac and non-cardiac complications of Kawasaki disease

Cardiac
Coronary artery aneurysm
Myocarditis
Valvulitis, usually mitral
Pericardial effusion
Systemic artery aneurysms
Myocardial infarction
Rupture of coronary aneurysm with haemopericardium
Non-cardiac
Arthralgia/arthritis
Aseptic meningitis
Hepatic dysfunction
Hydrops of gallbladder
Diarrhoea
Otitis media
Uveitis
Pneumonitis
Erythema and induration at BCG inoculation site
Peripheral gangrene

accompanied by abdominal pain and repeated vomiting of undigested food. She was initially admitted to a private hospital where she was noted to have jaundice, poor peripheral circulation and oliguria. She was then transferred to our PICU in the evening of the same day. The child was in shock on admission. Jaundice was evident. Erythematous maculopapular rash was seen over the trunk and the proximal part of the limbs, with the face relatively spared. Multiple lymph nodes were palpable on both sides of the neck, the largest one measured 1.5 cm in diameter. Conjunctival injection was not evident initially but became florid on the second day. Liver measured 3 cm below the right costal margin at the mid-clavicular line. The cardiac apex was displaced laterally. Initial laboratory investigation revealed gross leucocytosis (>40 x 10^9/l) with neutrophil predominance, normal platelet counts and hyperbilirubinaemia (160 μmol/l, 90% direct bilirubin). Alanine transferase (ALT) and gamma glutaryl transferase (γGT) were 2-4 times their respective upper limits. Creatine kinase (CK) and lactate dehydrogenase (LDH) were normal, while troponin I was elevated. Electrocardiogram showed nonspecific ST depression in leads II, and V2 through V5. Echocardiogram on the first day of admission revealed poor myocardial contactility with a shortening fraction of 19%. Ultrasound of the abdomen revealed a hydropic gallbladder and mild ascites. Thus the clinical picture was one of persistent pyrexia with exanthem, enlarged cervical lymph nodes and non-purulent conjunctivitis complicated by cardiogenic shock, hepatic impairment and hydrops of the gallbladder. Central venous pressure monitoring commenced (initial CVP read 24 mmHg). Dopamine and dobutamine infusions were given. Empirical broad-spectrum antibiotic cover was started. The most important differential diagnosis was fulminant viral infection (particularly adenovirus) and KD. Because of the high suspicion of KD, a dose of intravenous immunoglobulin (IVGG, 2 g/kg) was given on the second day of admission. Fever started to subside. Cracking of the lips was then noted and desquamation occurred first in the toes and perineum then in the fingers. Echocardiogram performed on the seventh day of admission (twelfth day since the onset of fever) revealed a coronary aneurysm of 4 mm in diameter in the middle segment of right coronary artery. Meanwhile, extensive investigations did not yield findings supporting alternative possibilities, the diagnosis of Kawasaki disease was established. A second dose of immunoglobulin was given in view of persistent low-grade fever. Platelet count and erythrocyte sedimentation rate steadily increased. Liver enzymes and bilirubin levels normalized after the first dose of IVGG. Aspirin was given at anti-inflammatory dose (30 mg/kg/day). Myocardial function improved gradually and inotropes were discontinued on the eighth day of admission. The child was discharged from PICU on the ninth day. Follow-up by paediatric cardiologist four weeks after onset of disease revealed persistent coronary arterial aneurysm. Myocardial function was regularly monitored and gradual normalization was documented (fractional shortening 33% three months after the acute phase). The gallbladder was not palpable and liver enzyme level stayed within normal limits. The child was maintained on antiplatelet dose of aspirin.

Discussion

Through the years there have been reports describing unusual presentation of KD mimicking sepsis with multi-organ involvement.[4,5] In our patient, KD disguised itself as fulminant sepsis with shock, renal shutdown and hyperbilirubinaemia. Cardiogenic shock secondary to myocarditis dominated the early phase of the illness when the cardinal features of KD did not fully manifested themselves yet. However, there were a few clinical features pertaining to KD, including non-purulent conjunctivitis, rash, lymph nodes and persistent fever, were present. Thus KD should be included in the differential diagnosis of a febrile toxic-looking child with multiorgan dysfunction.

Myocarditis is an important cardiovascular complication of Kawasaki disease besides coronary arterial aneurysm. It was reported in up to 50% of cases.[2] It can be seen during the first week after the onset of fever. It is most often manifested by tachycardia excessive for the degree of fever. ECG changes such as prolonged PR interval, ST-T segment changes and decreased voltage of R waves may occur. Deranged cardiac enzymes and raised troponin I have been documented, representing myocardial injury. Our patient illustrates the rare situation where myocarditis was severe enough to cause congestive heart failure and cardiogenic shock. Supportive care formed the integral part of the management. Close monitoring of vital signs, judicious fluid therapy and inotropic support were essential. The question whether immunoglobulin therapy has a direct positive effect on myocardial contractility remains unanswered, although one recent small study demonstrated a rapid improvement in myocardial contractility following immunoglobulin therapy.[6] In our case, the myocardial recovery seemed to be gradual, and no dramatic change was seen after each of the two doses of IVGG.

Hydrops of the gallbladder can be defined as acute distension of the gallbladder in the absence of mechanical obstruction to the cystic duct.[7] The diagnosis was considered when a right upper quadrant mass was palpable and/or seen on the plain radiograph. The ultrasonographic appearance is typically biconvex in longitudinal axis in contrast to the normal oval configuration.[8] The clinical conditions that may be associated with gallbladder hydrops are listed in Table 2.

Gallbladder hydrops is a well-known but uncommon complication of KD. Incidence rates quoted by early reports tend to be low. Bell et al[9] reported the rates of various complications of Kawasaki disease in the United States from July 1976 to December 1980. Gallbladder hydrops occurred in 2.9% of the 274 cases studied. A substantially higher incidence rate was reported by Suddleson from Los Angeles in 1987.[10] The series contained 232 cases, 117 of them underwent abdominal ultrasonography because of the presence of clinical findings of hepatomegaly, gallbladder enlargement, abdominal mass or jaundice. Sixteen (13.7%) children were found to have hydrops of the gallbladder. Abdominal pain (10/16) and vomiting (8/16) were the most prevalent symptoms. Hyperbilirubinaemia >20 μmol/l and palpable gallbladder were only found in three and four patients respectively. Of these 16 patients, fifteen of them were treated non-operatively with resolution of gallbladder hydrops occurring within 14 days in 80%. None of them had persistent hydrops beyond 60 days. No recognizable sequelae were seen. Only one case underwent cholecystectomy. The gallbladder histology revealed features of suppurative cholecystitis.

Table 2 Conditions associated with hydrops of the gallbladder

Neonate	
	Sepsis
	Hyperalimentation
	Inspissated bile syndrome
Children	
	Scarlet fever
	Kawasaki disease
	Leptospirosis
	Ascariasis
	Typhoid fever
	Familial Mediterranean fever
	Sepsis
	Total parenteral nutrition
	Polyarteritis nodosa

In our patient, gallbladder hydrops manifested as repeated emesis, jaundice and abdominal pain. These symptoms were nonspecific and occurred well before the full-blown emergence of the telltale features of KD. If jaundice were not present in this case and probably an ultrasound would not have been performed, then we would have missed the gallbladder hydrops. As mentioned above, hyperbilirubinaemia was only present in a minority of patients with gallbladder hydrops. It is probable that some hydropic gallbladders of Kawasaki patients may go unnoticed.

From reports dated back to the pre-IVGG era, it seems that most of the gallbladder hydrops complicating Kawasaki disease resolve spontaneously. In our case, a dramatic decrease of bilirubin level was noted after the first dose of immunoglobulin, but unfortunately we missed the chance of repeating the ultrasound at that juncture. Reports on gallbladder hydrops in the post-IVGG era are scarce, leaving the question whether immunoglobulin therapy can hasten the resolution of gallbladder hydrops unanswered.

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