Acute febrile cholestasis as an inaugural manifestation of Kawasaki’s disease

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ABSTRACT

We report a child who developed acute febrile cholestasis with jaundice and pruritus as the inaugural manifestation of Kawasaki’s disease (KD). The severe obstructive icterus and hydrops of the gallbladder required cholecystectomy that was not followed by remission of the fever and cholestasis. KD was suspected after the exclusion of all infectious, metabolic and neoplastic conditions responsible for acute cholestasis. The administration of intravenous gammaglobulin (IVGG) promptly induced defervescence and improvement of the patient’s general condition.

Mucocutaneous alterations, peeling of the digits, right cervical lymph node enlargement and bilateral non-suppurative conjunctivitis supporting the diagnosis of KD developed 14 days after the appearance of jaundice. No coronary abnormalities had developed after 2 years of follow-up. We conclude that this syndrome should be suspected in any child with febrile cholestasis of unknown origin, in order that coronary involvement may be prevented by the administration of IVGG.

Introduction

In the last decade atypical presentations of Kawasaki’s disease (KD) have been increasingly reported. The current therapy for KD, i.e. aspirin and intravenous gammaglobulin (IVGG), is also suggested in these patients in order to prevent the risk of coronary aneurysms (1).

In children with KD, hepatobiliary involvement is described as a minor clinical manifestation during the course of the disease. Hepatobiliary dysfunction, intrahepatic bile duct damage and hydrops of the gallbladder are frequently associated with acute KD, whereas jaundice or increased levels of bilirubin are rarely reported (2). Liver enlargement has been observed in 14.5% of patients (2) and a significant increase in aminotransferase levels has been reported in 20-30% of affected children (3).

We report a child who developed an acute febrile cholestasis as the initial clinical manifestation of KD.

Case report

A 2 year, 7 month old, previously healthy girl was admitted to the Paediatric Department with a 6-day history of high fever, arthralgia in her right lower limb, abdominal pain, itching jaundice, pale coloured stool and dark urine. Blood tests revealed a erythrocyte sedimentation rate (ESR) of 120 mm/hr, C reactive protein 18.25 mg/dL (n.v. < 0.5), white blood cells 24,400/ml with neutrophils 85%, lymphocytes 14% and eosinophils 1%, red blood cells (RBC) 3,550,000/ml, haemoglobin (Hb) 10 mg/dL, platelet count (PTL) 392,000/ml, aminotransferase ALT 89 U/L (n.v < 40), AST 62 U/L (n.v. < 40), total bilirubin 7.7 mg/dL with direct bilirubin 3 mg/dL.

Blood and urine cultures were negative and serological tests excluded Cytomegalovirus, Epstein Barr virus, and hepatitis A, B, and C virus infections. As her general condition rapidly deteriorated and the abdomen was tense and painful, a plain abdomen x-ray was performed which revealed liver enlargement and the diffuse presence of gas. Abdominal ultrasound showed a markedly distended gallbladder with anechoic content. The intrahepatic ducts were enlarged while the common bile duct was normal. Due to the severity of the abdomen distension, laparatomy was performed which showed severe hydrops of the gallbladder.

Given the high risk of rupture, the patient underwent cholecystectomy including the resection of her Mascagni’s lymph node. Histology of a liver biopsy showed inflammatory infiltrates in the portal tract while liver architecture was preserved. The inflammatory infiltrates, consisting of eosinophils and polymorphonuclear leukocytes, were localised around and within the lumina of the bile ducts. A bile culture was negative. Despite cholecystectomy and wide-spectrum antibiotic treatment, high fever and severe jaundice persisted. All the markers of inflammation and cholestasis were still raised, while RBC decreased to 2,890,000/ml with Hb 8.2 g/dL and PTL increased up to 1,080,000/ml.

After the exclusion of all known infectious, metabolic and neoplastic diseases responsible for acute cholestasis, the association of high fever recalcitrant to an-
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Since intrahepatic cholestasis may benefit from hydrophilic bile ursodeoxycholic acid, this treatment was administered with a complete normalisation of the parameters of the biliary system over 2 months. Our case further supports the notion that in all children with acute febrile cholestasis of unknown origin, the diagnosis of KD should be suspected and IVGG therapy considered in order to prevent coronary involvement (11).

References