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KAWASAKI DISEASE ASSOCIATED WITH GALLBLADDER HYDROPS*

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SUMMARY: Coşkun Y, Bayraktaroğlu Z, Gökalp A, Çil A, Özkutlu S. (Departments of Pediatrics and Surgery, Gaziantep University Faculty of Medicine, Gaziantep, and Cardiology Unit, Department of Pediatrics, Hacettepe University Faculty of Medicine, Ankara, Turkey). Kawasaki disease associated with gallbladder hydrops. Turk J Pediatr 1995; 37: 269-273.

Kawasaki disease is an acute, febrile, systemic syndrome of unknown etiology which principally affects young children. We present a five-year-old boy, who developed an unusual finding, acute hydrops of the gallbladder, in addition to the characteristic signs and symptoms of Kawasaki disease. Clinical features of the disease and treatment regimens, including early administration of intravenous gamma globulin (IVGG), which prevents cardiac complications, are discussed. *Key words: Kawasaki disease, gallbladder hydrops.*

Kawasaki disease (KD), also known as mucocutaneous lymph node syndrome, is an acute, febrile, systemic syndrome of unknown etiology seen in early childhood. Although diagnostic tests for KD are lacking, the diagnosis of the disease is not difficult in patients with full expression of clinical features of the disease. The main clinical features of KD are: 1. fever lasting more than five days and unresponsive to antibiotics; 2. bilateral conjunctival hyperemia; 3. nonsuppurative cervical lymphadenopathy; 4. a non-vesicular exanthem, usually truncal; 5. one or more signs of oropharyngeal mucosa, including "strawberry tongue", pharyngeal hyperemia, and dry, fissured or injected lips; and 6. erythema and desquamation of the hands and feet[1].

This syndrome has rarely been reported in Turkey[2,3]. Here we present an unusual case of KD associated with gallbladder hydrops.

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Case Report

A five-year-old boy was admitted to the pediatric department of Gaziantep University Hospital with the complaints of a three-day history of fever, cervical bilateral lymphadenopathy, rash, abdominal pain, vomiting, dark urine and jaundice (Fig. 1a).

Clinical examination revealed a well-developed child with a body weight of 18 kg. His body temperature was 40 °C. The main physical findings were: diffuse erythematous maculopapular rash, bilateral cervical lymphadenopathy a diameter of with 3 cm; dry, red, fissured lips; "strawberry tongue"; hyperemia in the pharynx; bilateral hyperemic conjunctivitis; icteric sclera; abdominal discomfort and irritability with conspicuous neck stiffness; and Kernig and Brudzenski signs (Fig. 1b). Laboratory investigations initially revealed a hemoglobin level of 7.4 g/dl and white blood count of 16,600/mm^3, with a distribution of 81% segmented neutrophis, 5% juvenile neutrophils, 2% monocytes, 12% lymphocytes and 305,000/mm^3 platelets, The sedimentation rate was 120 mm/h. Urine analysis showed marked bilirubinuria and trace proteinuria. Blood electrolytes, BUN and creatinine were within normal limits. Liver function tests were abnormal: total protein 45 g/L, albumin 25 g/L, total bilirubin 122.4 µmol/L, direct bilirubin 95.2 µmol/L, alkaline phosphatase 49.6 nmol/L, ALT 1.1 µmol/L and AST 1.3 µmol/L.

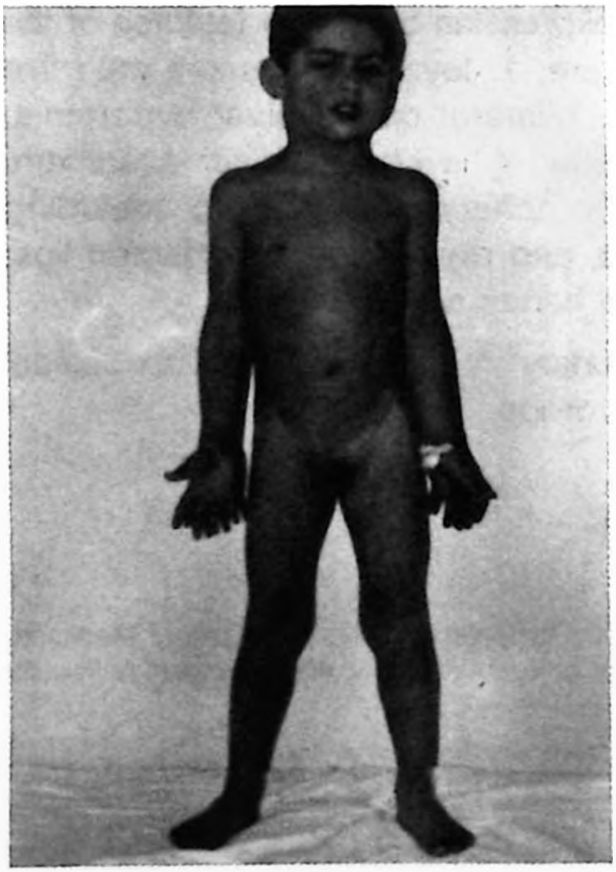

Fig. 1a: Non-vesicular exanthem and cervical lymphadenopathy.

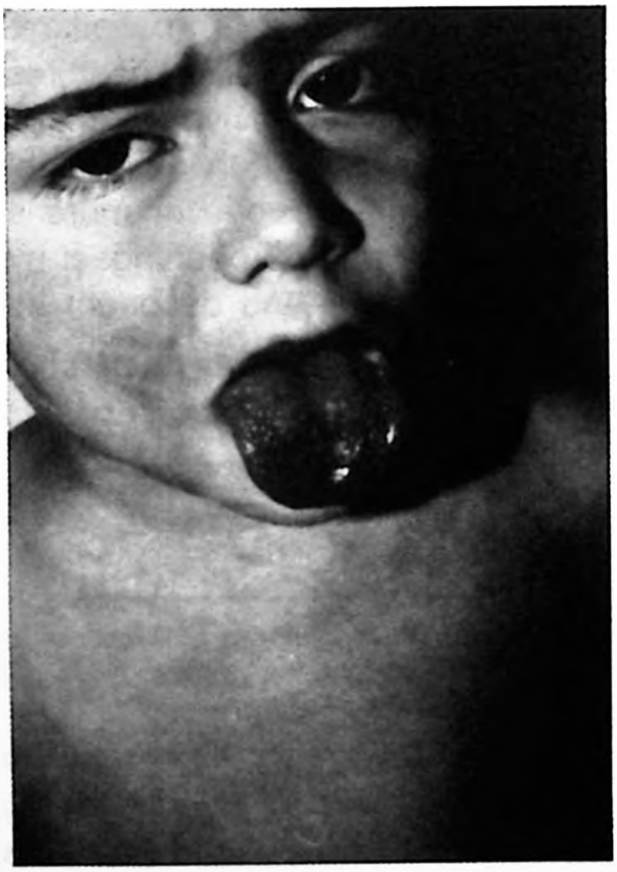

Fig. 1b: Injected lips and strawberry tongue of the patient.

Other laboratory studies, including blood, throat and feces cultures, Brucella and Salmonella agglutination tests, HBsAg and anti-HBsAg, serum immunoglobulin levels (IgG, IgM, IgA, IgE), C3, C4, RF and ASO titers, were negative or within normal limits. Hydropic gallbladder was discovered by abdominal ultrasonography (USG) (Fig. 2). Cardiomegaly was not detected on chest x-ray. At first, electrocardiogram (EKG) revealed no abnormality except sinus tachycardia. Echocardiographic examination in the first and eleventh months were normal.

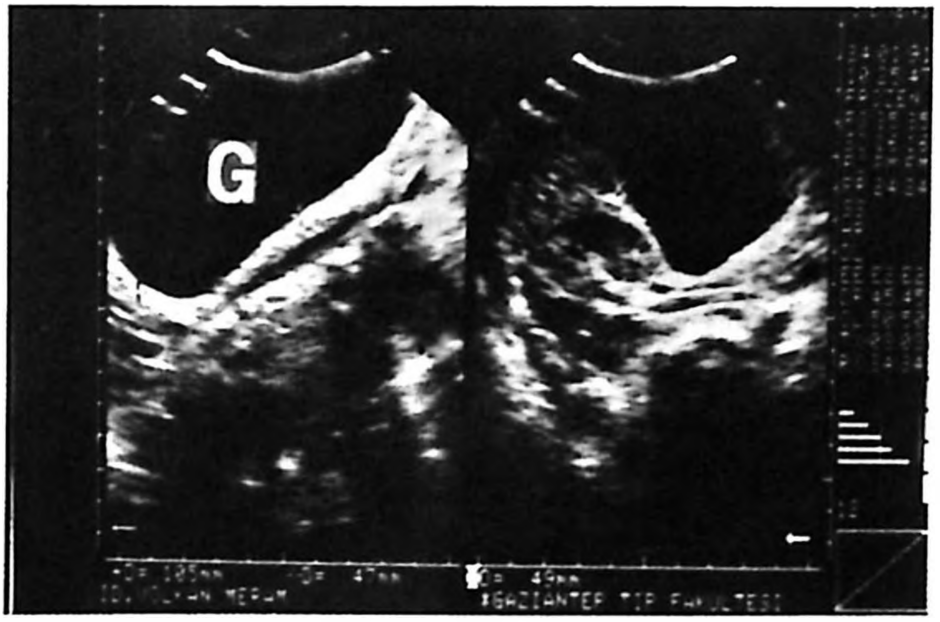

Fig. 2: The appearance of hydropic gallbladder in ultrasonography. G: gallbladder.

On the first day of hospitalization, intravenous (IV) potassium penicillin G administration was initiated at 300,000 units/kg/24h in six divided doses. Then on the fourth day, administration of penicillin G was stopped and ceftriaxone at a dose of 2 g/24h was started and given for ten days. On the second day, in addition to antibiotherapy, acetylsalycylic acid at a dose of 100 mg/kg24h and IV immunoglobulin (400 mg/kg/24h) were commenced and given for three weeks and seven days, respectively. The patient had daily fevers as high as 39-40 °C for ten days. The rash gradually increased, and then desquamation was observed mainly on the trunk, spreading to the extremities; in addition, marked abdominal pain and discomfort continued during the first month of the disease.

The hydrops of the gallbladder recovered according to ultrasound findings, and serum bilirubin levels returned to normal at the end of the fourth week. At the beginning of the second week of the disease, thrombocytosis up to one million/mm^3 and slight ST depression and prolonged PR interval on EKG were detected; thus, dipyridamole 3 mg/kg/24h was added as an anticoagulant therapy and continued until the end of the second month when thrombocytosis subsided. The patient received acetylsalycylic acid at a dose of 30 mg/kg/24h for one month and then 5 mg/kg/24h for approximately one year.

Discussion

In our patient we observed high fever lasting ten days; bilateral cervical lymphadenopathy; dry, red, fissured lips; "strawberry tongue"; pharyngeal hyperemia; hyperemic conjunctivitis; diffuse maculopapular rash and desquamation on the trunk and extremities. With these well-known diagnostic criteria, the diagnosis of KD was not difficult to make, and the typical clinical course of the patient confirmed our diagnosis.

Although meningeal signs are uncommon in KD they may be seen as in our patient, suggesting central nervous system involvement. The other main finding in the patient was jaundice. This finding is not usual in KD, although mild abdominal pain and discomfort are occasionally seen in KD as a sign of hepatic involvement. Marked abdominal pain particularly in the right upper quadrant of the abdomen and associated with jaundice in our case suggested a hepatobiliary complication of Kawasaki syndrome[4]. Then, in the ultrasound examination we detected profound hydropic gallbladder, which is an unusual finding of the disease. Hydrops of the gallbladder did not resolve before one month, and the patient had slight abdominal discomfort lasting for several months. The cause and incidence of this rare complication is not exactly known. Some explanations for the occurrence of gallbladder hydrops, such as regional lymph node compression in the porta hepaticus, have not been well documented[5].

Electrocardiographic changes and cardiac omplications are commonly seen in KD, with frequencies of 74 percent and 25-30 percent, respectively[5-7]. We found PR prolongation and slight ST depression on EKG with no abnormal finding in echocardiography in the acute phase of the disease. The EKG changes resolved within two months, and no abnormality was demonstrated by echocardiography in long-term follow-up (12 months). The other main feature that occurs in almost all patients is thrombocytosis that usually appears in the second week of the disease; thus, management of platelet aggregation is an important issue for decreasing the risk of cardiovascular complications.

Previous reports have strongly demonstrated that early administration of high dose intravenous gammaglobulin (IVGG) plus acetylsalycylic acid will prevent cardiac complications of this syndrome[8]. We started administering IVGG plus acetylsalycylic acid on the second day of hospitalization and added dipyridamole at the end of the second week when the thrombocytosis was profound.

Prevention of cardiac complications was most likely due to early administration of IVGG and long-term use of acetylsalycylic acid and dipyridamole in the thrombocytotic state. Whether IVGG administration was effective in the early resolution of gallbladder hydrops is not clear.

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