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GALLBLADDER HYDROPS IN AN INFANT WITH KAWASAKI DISEASE

Case Report

KAWASAKİ HASTALIĞI OLAN BİR BEBEKTE SAFRA KESESİ HİDROPSU

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ABSTRACT

Kawasaki disease is the leading cause of acquired heart disease in childhood. Gallbladder hydrops is an uncommon manifestation of this disease, found rarely in infants. We report the case of Kawasaki disease in a 9-month-old boy with cardiac involvement and abdominal distention related to gallbladder hydrops which was diagnosed by ultrasonography and resolved spontaneously during follow-up.

Key words: *Gallbladder hydrops; infant; Kawasaki disease* .

ÖZET

Kawasaki hastalığı çocukluk çağındaki kazanılmış kalp hastalığı nedenlerinden biridir. Safra kesesi hidropsu bu hastalığın seyrek görülen bulgularından olup, bebeklerde nadirdir. Abdominal distansiyonun safra kesesi hidropsuna bağlı olduğu ultrasonografiyle gösterilen ve takibinde hidrops bulguları gerileyen 9 aylık ve kardiyak bulgusu olan Kawasaki hastalığı olgusu sunulmuştur.

Anahtar kelimeler: *Bebek; Kawasaki hastalığı; safra kesesi hidropsu.*

INTRODUCTION

Mucocutaneous lymph node syndrome or Kawasaki disease (KD) is an acute, febrile, multi-systemic illness associated with multiorgan vasculitis of unknown etiology, first reported by Kawasaki et al.1 Because of the absence of specific diagnostic tests and unknown etiology and pathophysiology, physicians must rely upon the presence of specific clinical criteria and laboratory data that support the diagnosis of KD. Prolonged fever (characteristically high, remittent), conjunctivitis (bilateral bulbar conjunctival injection, usually without exudate), oropharyngeal hyperemia (erythema of the oral and pharyngeal mucosa with strawberry tongue and dry, cracked lips, and without ulceration), widespread rash of various forms (maculopapular,

erythema multiforme, or scarlatiniform), lymphadenopathy (nonsuppurative cervical lymphadenopathy, usually unilateral, with node size of ≥1.5 cm), edema, and hyperemia of the extremity and desquamation are common clinical findings of KD. Typical KD requires a high fever for >5 days and at least 4 of 5 other clinical manifestations. Most children present with symptoms by the age of 5 years. Hydrops of gallbladder is less common, occurring in 15% patients in the first two weeks of the disease which most commonly affects the children aged from 17 months to 7 years.2,3,4 We report the case of KD in which hydrops of the gallbladder was associated with abdominal pain and was diagnosed by ultrasonography.

CASE REPORT

A 9-month-old boy was admitted to our emergency department with a 6-day history of fever and reluctance to food. On physical examination, he was irritable with a temperature of 39^{0}C, respiration rate of 44/min, and heart rate of 158 beats/min. Oral mucosa and tongue were hyperemic; he also had bilateral bulbar conjunctival injection without exudate, a maculopapular rash on his chest and abdomen, and his BCG scar was flushed. Laboratory studies showed neutrophilic leukocytosis (17600/mm^3, normal range: 6–17500/mm^3) and an elevated erythrocyte sedimentation rate (95 mm/h, normal range: 0–20 mm/h) and C-reactive protein (257.3 mg/dL, normal range: <2.8), blood urea nitrogen (BUN) and creatinin were slightly high (BUN: 31 mg/dL, normal range: 4-19, creatinin 0.44 mg/dL, normal range 0.17—0.42), γ-glutamyl transpeptidase (GGTP) level was found high (172 U/L, normal range <32) and pyuria was seen in urine analysis. He was hospitalized with the suspicion of KD based on the clinical findings. Echocardiographic investigation showed hyperechogenicity of the pericoronary tissue and large left anterior descending artery-left circumflex artery. No aneurysm was detected. Blood, urine and stool cultures were all negative. In the first two days of therapy, he received 1 g/ kg intravenous immunoglobulin G (IVIG) with 100 mg/kg aspirin. On the 3rd day, right upper quadrant tenderness and a palpable mass was revealed on physical examination. A sonogram showed gallbladder hydrops with a diameter of 8.8 cm (**Figure 1**).

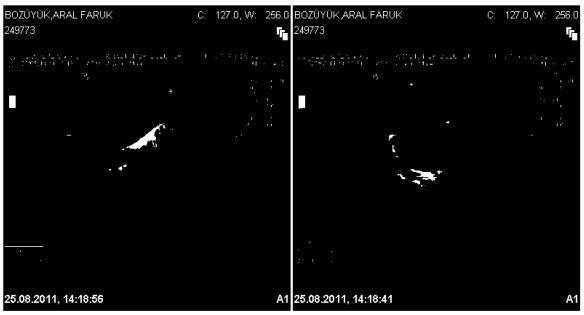

Figure 1. *Gallbladder hydrops (On the 3rd day). There is no internal wall irregularity. Intraluminal echo and wall thickness (<2 mm) are normal. Gallbladder dimensions are 84x69 mm.*

During follow-up, the acute-phase reactants became normal. Elevation in his GGTP test result as follows 89 U/L and troponin I less than 0.02 ng/ml. Serial sonograms were obtained for following the resolution of the hydrops. Supportive medical therapy was started and oral feeding was stopped. Symptoms of gallbladder hydrops subsided within one week of treatment. During follow-up, ultrasonography revealed a slower regression of the gallbladder hydrops.The periodic measurements of the diameter and thickness of the gallbladder wall are shown in **Table 1**.

Days	Gallbladder diameter in lenght (mm)	Gallbladder diameter in width (mm)	Gallbladder wall thickness	Pericholecystic fluid.
3	84	69	<2 mm	Absent
5	88	44	<2 mm	Minimal
6	72	40	<2 mm	Minimal
12	30	43	<2 mm	Absent

Table 1. *Periodic radiologic findings of the gallbladder determined by ultrasonography.*

There was no complication necessitating surgical intervention and, 12 days after his initial presentation the ultrasonographic appearance of the gallbladder was normal (**Figure 2**).

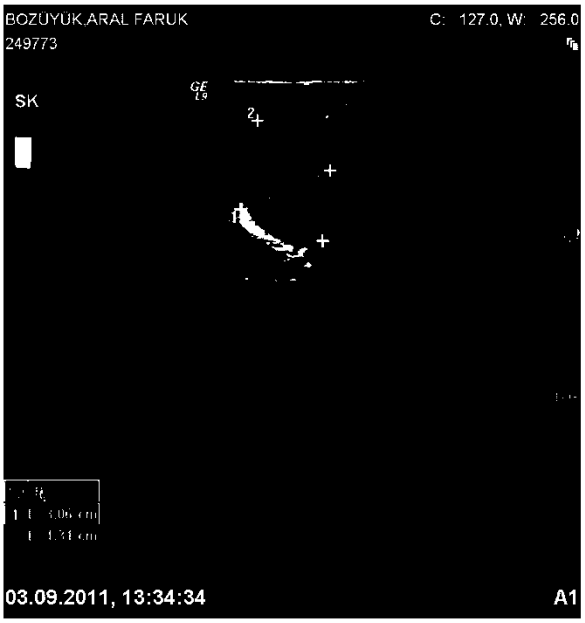

Figure 2. *Finally, 12 days after his initial presentation the ultrasonographic appearance of the gallbladder was normal. There was no pericholecystic fluid. Gallbladder dimensions are normal (43x30 mm).*

And 14 days after hospital admission, his echocardiographic examination became normal. The patient is being followed up closely for signs of any late complications of KD.

DISCUSSION

Mucocutaneous lymph node syndrome or KD was first described by Tamisaku Kawasaki in 1967. Despite extensive investigation, the etiology of KD remains unknown, so the diagnosis depends on non-spesific clinical signs rather than a definitive laboratory test. In addition to the diagnostic criteria, a broad range of non-spesific clinical features are associated with KD, including abdominal pain, arthralgia, aseptic meningitis, cough, diarrhea, gallbladder hydrops, hepatitis, irritability, lethargy, parotitis, rhinorrhea, seizure, semicoma, uretritis, uveitis and vomiting.(5,6,7,8)Prolonged fever, bilateral bulbar conjunctival injection without exudate, hyperemic changes on lips and in the oral cavity, widespread polymorphous exanthema, cervical lymphadenopathy, edema, and hyperemia on the extremities and desquamation are common clinical findings of KD. (3) The cardiovascular involvement is the major determinant for morbidity and mortality. (9).

The gastrointestinal symptoms of KD include abdominal pain, diarrhea, distention, cholestatic jaundice and vomiting. (2). Gallbladder hydrops is a rare manifestation of KD and occurs in only 3%–12.7% of all patients. it is being recognized more frequently with improved ultrasonography because of its well-known association with KD. (10). KD usually affects children between the ages of 3 months and 12 years; however, gallbladder hydrops seems to occur between the ages of 17 months and 7 years. (11). KD with gallbladder hydrops occurred at a mean age of 5.2 years. (12). In the view of literature our patient is one of the youngest case of KD with gallbladder hydrops.

The manifestations of gallbladder hydrops occurring with KD were abdominal pain (100%), right upper quadrant tenderness (90%), vomiting (75%), and a palpable mass (55%). (6).Diagnosis is established by ultrasonography of the abdomen demonstrating normal biliary ducts and a distended gallbladder withsonalucent appearance and spherical configuration without calculi or congenital malformation. A review of the literature reveals a study documenting gallbladder size in normal childrenwhich are documented in **Table 2**. (14).

Age (Years)	Mean (mm)	Range (mm)
0-1	25	13-34
2-5	42	29-52
6-8	56	44-74
9-11	50	34-65
12-16	61	38-80

Table 2. *Lenght of gallbladder in normal children determined by ultrasonography (McGahan et al).*

The mean duration of gallbladder hydrops associated with KD is 15 days.The resolution can be taken up to 60 days. (6) In our patient, the gallbladder did not appear normal until 12 daysafter the initial signs.

The aetiology of acute hydrops of the gallbladder in KDmay be multifactorial. Hypertrophied inflamed nodes may cause reactive inflammation and obstruction of the cystic duct, which probably results in acalculous distention of the gallbladder.[16] Gallbladder hydrops may be exacerbated by prolonged fasting, fever and dehydration, with consequent bile stasisanditmay be related to nonspecific vasculitis.[18] Inefficient immune response to vasculitis may cause, prolonged resolution of gallbladder hydrops.Also gallbladder hydrops in children has been associated with gastroenteritis, viral hepatitis, leptospirosis, upper respiratory tract infections, staphylococcal abscess, familial Mediterranean fever, scarlet fever, polyarteritis nodosa, leukemia and nephrotic syndrome. (12,6,19).

As in our patient, the clinical course of gallbladder hydrops associated with KD is almost always benign and self-limited. The management of gallbladder hydrops in KD is nonoperative and it can be treated symptomatically. (11,20,21) Supportive treatment with intravenous fluids, analgesics, and gastrointestinal rest is indicated. (11). The role of antiinflamatuar drugs are unclear. If gallbladder hydrops is caused by vasculitis, salicylate therapy may be effective in prevention if treatment is begun early in the course of illness. Corticosteroids may be contraindicated because of an increased risk of aneurysm formation.(22) Surgical intervention (cholecystostomy) should be done only if the child is unresponsive to medical therapy or if there are signs of bile peritonitis secondary to gallbladder perforation. (11) The patient should be followed up with repetitive clinical examinations and serial sonograms.[6,12] Ultrasound is the optimal method for evaluating these patients. Most cases resolve within one to two weeks of diagnosis, andcomplete resolution may extend up to four weeks. (12).

In conclusion, gallbladder hydrops associated with KD is selflimited and may be seen in infants as young as 9 months old. Abdominal symptoms (e.g. abdominal pain, right upper quadrant tenderness and distention or vomiting) developing in a child with KD, the possibility of gallbladder hydrops should be entertained and the diagnosis should be confirmed by ultrasonography, which shows increased dimensions of gallbladder. Ultrasonographic evaluation and a high index of suspicion are mandatory for the early diagnosis.

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