Journal of Pediatric Gastroenterology and Nutrition
35:363–365 © September 2002 Lippincott Williams & Wilkins, Inc., Philadelphia

Case Report

Late Cholangitis After Kasai Procedure Detected With Magnetic Resonance Cholangiopancreaticography: A Case Report

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INTRODUCTION

Ascending cholangitis occurs in about 30 to 50% of patients after Kasai hepatoportoenterostomy for extrahepatic biliary atresia (1,2). It generally occurs within the first few years after surgery (3). Late cholangitis seems to be extremely rare (3,4). Patients with cholangitis usually have high fever with chills and jaundice, reflecting worsening of liver functions in about half of the patients (5). Blood tests may show leucocytosis. Diagnosis is best made by liver biopsy, showing histologic features of cholangitis. In addition, cultures of liver tissue and blood may be positive. Ultrasound and computer tomography (CT) scan usually reveal distended intrahepatic bile ducts. Postoperative ascending cholangitis is an important prognostic factor and is associated with liver cirrhosis, portal hypertension, and premature death. Because of the risk of permanent liver damage, aggressive therapy is fully justified.

We report the case of a girl who underwent a Kasai procedure at the age of 8 weeks and had a fully uneventful follow-up with normal liver functions in the following years. She was considered cured until the age of 8 years when she had a first episode of fever, tentatively diagnosed as cholangitis.

We were only able to actually diagnose cholangitis when magnetic resonance cholangiopancreaticography (MRCP) showed abnormalities consistent with sclerosing cholangitis during the fourth episode, which occurred when the patient was 17 years old.

CASE REPORT

An 8-week-old girl was diagnosed with extrahepatic biliary atresia and successfully underwent hepatoportoenterostomy according to Kasai. Liver functions normalized completely within 3 weeks. At follow-up, growth and development were normal and no adverse events occurred. As she appeared to be cured, follow-up was stopped when she was about 5 years old. At 8 years of age, however, she was seen again because of an episode of fever up to 39.5°C and pain in the right upper-abdominal quadrant. The pain worsened with coughing, breathing, and moving. She complained of nausea and vomited once. There was no jaundice. Micturition and defecation remained normal. At admission, serum aminotransferases were normal, but they reached peaks of three times the upper limits of normal after a few days (Fig. 1). There was marked leucocytosis. No infectious focus could be detected and empirical treatment was instituted with broad-spectrum antibiotics, including piperacillin and gentamycin. Blood cultures remained negative, liver biopsy did not present any signs of inflammation, and ultrasound was indecisive. Fever and pain disappeared and liver functions normalized. She was given prophylactic treatment with amphotericin B and cholistine for three months. She remained well for another 9 years.

At the age of 17 years she was admitted because of a similar episode with high fever and acute pain in the upper right abdomen. Although no objective signs of liver involvement were present, the same therapy was given, followed by oral ciprofloxacin for another 4 weeks. Ten days after she stopped taking oral ciprofloxacin, she had an identical third episode of illness. Liver biopsy again did not show any abnormality and repeated blood cultures were negative. With intravenous treatment with piperacillin and gentamycin for 2 weeks, the fever disappeared and she was given ciprofloxacin orally for 3 months.

A few weeks after discontinuation of prophylaxis, however, symptoms reappeared and it was decided to start a search for any possible other source before giving antibiotics. CT scan and ultrasound both failed to demonstrate intra-abdominal pathology, including the liver.

Received July 20, 2001; accepted April 17, 2002.

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DOI: 10.1097/01.MPG.0000024561.65871.21

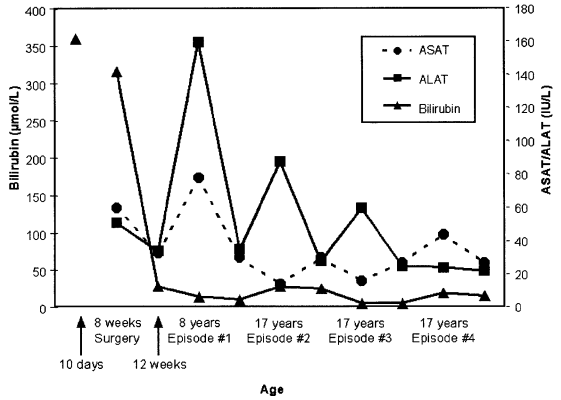

FIG. 1. Evolution of serum bilirubin and aspartate and alanine aminotransferases at time of surgery, during attacks of cholangitis and in between such attacks.

Gallium scintigraphy also was negative. MRCP, however, showed typical changes with abnormal bile ducts throughout the left lobe and the quadrate lobe, extending into the right lobe (Fig. 2). During this episode, therapy was postponed until the full diagnostic workup was completed. Finally a blood culture was obtained, which was positive for *Klebsiella pneumoniae*. She was intravenously treated with piperacillin, metronidazole, and gentamycin for 2 weeks. Liver functions normalized rapidly. As curative lobectomy was considered impossible, long-term oral prophylaxis with ciprofloxacin was instituted. At the age of 20 years, however, she had a fifth episode of cholangitis-related fever, again treated with piperacillin. Repeat MRCP confirmed the lesions represented in Figure 2. Presently she is under consideration for liver transplantation.

DISCUSSION

Extrahepatic biliary atresia is an obliterative cholangiopathy that involves all or part of the extrahepatic biliary tree and in many cases also the intrahepatic bile ducts (6). It has a worldwide incidence of approximately one in 10,000 live births (7). When left untreated, the average survival time is reported to be 19 months and the 3-year survival rate for children without any drainage procedure is less than 10% (6,8). Presently, 10-year survival without liver transplantation after hepatic portoenterostomy according to Kasai is reported to be 25 to 35% (7,9).

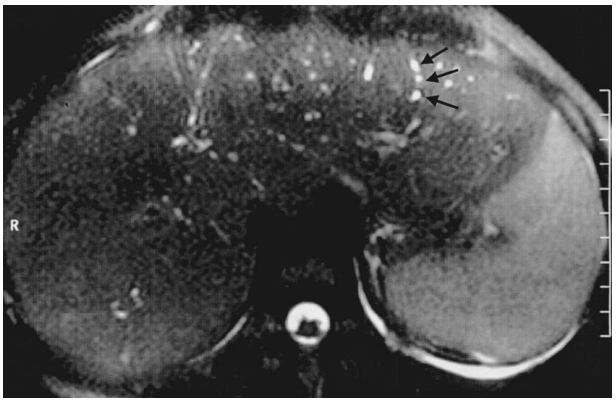

FIG. 2. MRCP of liver at age 17 years. Bile ducts appear irregular with distensions (arrows) separated by narrowings, mainly in the left liver lobe, indicative of (secondary) sclerosing cholangitis. Scale is in centimeters.

Some authors favor transplantation as the primary therapy (10). However, considering the shortage of infant donors and the direct relationship between age at transplantation and rate of survival, Kasai operation still remains the primary treatment for biliary atresia (9–13).

Ascending cholangitis is the most important postoperative complication. It has a strongly negative influence on prognosis after successful surgical repair, being the cause of progressive cirrhosis and death in a considerable number of cases (9,14–20). Patients who never presented with cholangitis have better prognosis than patients who presented with cholangitis; repeated attacks of cholangitis further decrease survival (14). Early detection and aggressive treatment of ascending cholangitis, therefore, is of great importance. Importantly, ascending cholangitis typically is a problem of the first few years of life (14,16,19); late occurrences seem to be rare (21,22).

We present the case of an adolescent girl with repeated episodes of unexplained fever, the first one 8 years after she successfully underwent a Kasai procedure and again at and after the age of 17 years. She was treated as having cholangitis, although the diagnosis could not be proven with conventional methods including cultures, liver biopsy, ultrasound, and CT scan. During those episodes, liver functions were only marginally and temporarily abnormal and liver biopsy revealed no signs of acute damage or cirrhosis. Only when MRCP was performed were typical signs of bile duct distension and narrowing detected in parts of the liver and interpreted as consistent with sclerosing cholangitis secondary to chronic infection. MRCP is only recently introduced as a noninvasive diagnostic modality for biliary tract disease (23,24). Unlike endoscopic cholangiopancreaticography and percutaneous transhepatic cholangiography, the procedure is noninvasive and free of complications such as pancreatitis, perforation, cholangitis, sepsis, bile leakage, and intraperitoneal hemorrhage (13).

Because MRCP creates high contrast between the hyperintense signal of bile and the low signal of the surrounding structures, it can provide complete visualization of the biliary tree. MRCP has excellent diagnostic accuracy in the diagnosis of biliary disease, fully comparable to invasive cholangiography (25) and may be even superior in the evaluation of sclerosing cholangitis (26).

To our knowledge, this is the first report on MRCP as a means of diagnosing late cholangitis after surgical intervention for biliary atresia. This case history suggests a

role for MRCP in the follow-up of patients with extrahepatic biliary atresia who are suspected of having hepatic pathology despite negative liver histology and cultures.

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