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Restrictive Cardiomyopathy in a Patient with Extrahepatic Biliary Atresia

The most commonly associated anomalies in patients with extrahepatic biliary atresia are cardiovascular, digestive and splenic defects. Of the cardiovascular anomalies, there are very few reports of biliary atresia with cardiomyopathy. We report the first case of a child with extrahepatic biliary atresia and restrictive cardiomyopathy. The patient was a 13-month-old boy diagnosed with extrahepatic biliary atresia at the age of 2 months, when he underwent laparotomy for definite diagnosis. Hepatic portoenterostomy was performed after confirmative cholangiogram. Recently, he developed severe cough and dyspnea, and his respiratory symptoms worsened. Chest radiograph showed cardiomegaly. Two-dimensional echocardiography showed marked biatrial enlargement. On M-mode echocardiogram, a slight increase in left ventricular dimension was seen in early diastole with a relatively good left ventricular function. Mitral inflow Doppler tracing showed an increased E-velocity (1.1 m/sec) with decreased deceleration time (75 m/sec), and increased E/A ratio (0.33). He was diagnosed as having restrictive cardiomyopathy with characteristic echocardiographic features.

Key Words : *Bile Ducts, Extrahepatic; Biliary Atresia; Cardiomyopathy, Restrictive*

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INTRODUCTION

Extrahepatic biliary atresia is due to a process of unknown etiology that leads to the complete obstruction or disappearance of extrahepatic bile ducts in infants. It affects 1 in 1,500 live births (1, 2). The cause of the anomaly, pathogenic mechanisms, and moment of production of the lesion are unknown. The incidence of associated malformations in patients with extrahepatic biliary atresia is as high as 25% (3). The most commonly reported anomalies are cardiovascular, digestive and splenic defects (4).

Of the cardiovascular anomalies, there has been a report on dilated cardiomyopathy associated with biliary atresia (5). To our knowledge, however, none of the reported cases with biliary atresia had restrictive cardiomyopathy. We report the first case of a child with extrahepatic biliary atresia and restrictive cardiomyopathy.

CASE REPORT

The patient was a 13-month-old boy diagnosed with extrahepatic biliary atresia at the age of 2 months, when he underwent laparotomy for definite diagnosis. Hepatic portoenterostomy was performed simultaneously after confirmative cholangiogram. There was no other associated anomaly such as polysplenism. Liver biopsy showed that there were bile ductular proliferation and the presence of bile plugs, with the basic hepatic lobular architecture intact. Mild cardiomegaly shown at the initial chest radiography was neglected because of the absence of cardiac symptoms. Subsequently, he has had several episodes of ascending cholangitis and intermittent respiratory difficulties. In the last few days before this admission to our hospital, his respiratory symptoms including cough and dyspnea worsened. He developed cyanosis and general weakness, and was thus brought to the hospital.

On physical examination he was pale and dyspneic. He weighed 7.5 kg and his height was 69 cm (both were below the 3rd percentile). His temperature was 38.4℃, pulse 122/min, blood pressure 80/50 mmHg, and respiration 22/min. His cardiac examination revealed a quiet precordium, regular rhythm and normal heart sound, and his chest was clear to auscultation bilaterally. The abdomen was markedly distended and the liver was palpable at 4 cm and spleen at 5 cm below the costal edge. The extremities were edematous and cyanotic.

Laboratory data were as follows: White blood cell

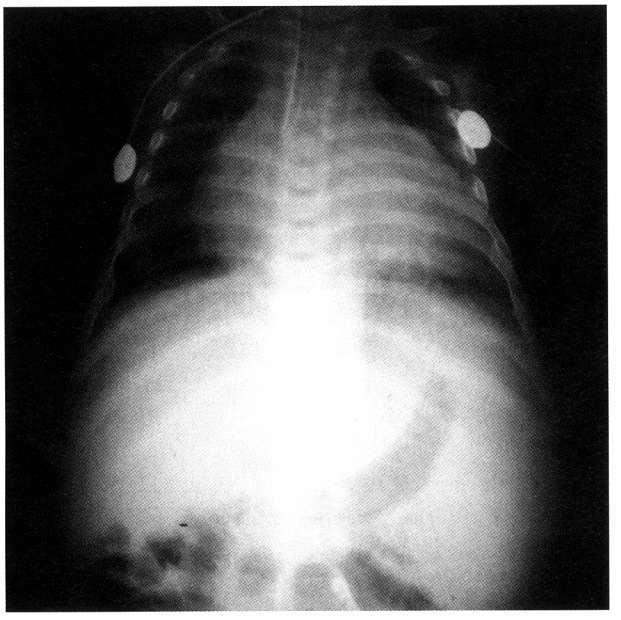

Fig. 1. Chest radiograph shows cardiomegaly.

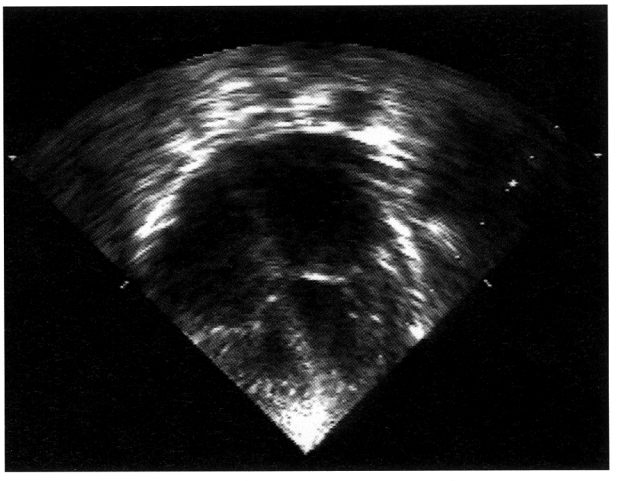

Fig. 2. Apical four-chamber view of two-dimensional echocardiography shows marked biatrial enlargement.

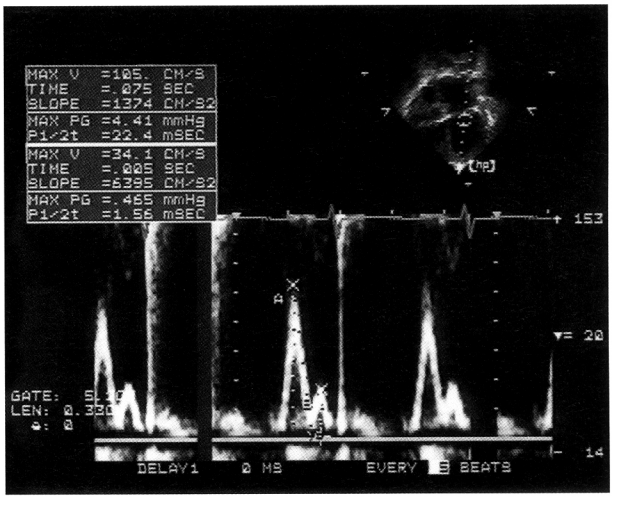

Fig. 3. Mitral valve Doppler recording from the patient.

17,200/mL (69% neutrophils), platelets 311,000/μL and hemoglobin 6.6 g/dL. Total bilirubin was 1.7 mg/dL, direct bilirubin 0.9 mg/dL, aspartate aminotransferase 52 IU/L, alanine aminotransferase 24 IU/L, akaline phosphatase 452 IU/L, total protein 7.2 g/dL, albumin 4.0 g/dL and total cholesterol 115 mg/dL. Coagulation and serum electrolyte levels were normal. Arterial blood gas determination revealed a metabolic acidosis: pH 7.27, $PaCO_2$ 30, PaO_2 57, HCO_3 13, and -12 base deficit.

Chest radiograph showed cardiomegaly (Fig. 1). The electrocardiogram showed prominent P waves and normal QRS voltage. Two-dimensional echocardiography showed marked biatrial enlargement (Fig. 2). On M-mode echocardiogram, a slight increase in left ventricular dimension was seen in the early diastole with a relatively good left ventricular function. Mitral inflow Doppler tracing showed an increased E-velocity (1.1 m/sec) with decreased deceleration time (75 m/sec) and increased E/A ratio (0.33). LV end-systolic dimension was 2.14 cm, and LV end-diastolic dimension, 3.36 cm (Fig. 3).

He was diagnosed as having restrictive cardiomyopathy with characteristic echocardiographic features. Treatment was directed toward the relief of edema with diuretics. He has since fared relatively well although chest radiograph still shows cadiomegaly. He is four years old and is being followed up at regular intervals.

DISCUSSION

Biliary atresia is the most common cause of extrahepatic obstructive jaundice in the newborn and is the most frequent indication for liver transplantation in children (6). Clinically, biliary atresia occurs as an isolated abnormality or in combination with other congenital anomalies. Ten to thirty percent of cases are associated with developmental abnormalities of the heart, digestive tract, or spleen or other anomalies such as situs inversus, malrotation, preduodenal portal vein and absent inferior vena cava (7, 8). Silveira et al. observed that 20% of children with extrahepatic biliary atresia had significant associated anomalies, and in these patients 60% had cardivascular abnormalities (4). They were pulmonary stenosis, ventricular septal defect, patent ductus arteriosus, atrioventricular septal defect, atrial septal defect, dextrocardia, coarctation of aorta, oval fossa defect, right juxtaposition of atrial appendages, hypoplasia of left heart and aortic stenosis.

However, there are very few reports of biliary atresia

with cardiomyopathy. Especially restrictive cardiomyopathy associated with extrahepatic biliary atresia has never been reported to our knowledge. The echocardiographic findings were consistent with restrictive cardiomyopathy in our patient. The most striking two-dimensional echocardiographic feature of restrictive cardiomyopathy is the enormous biatrial enlargement seen in the apical and subcostal four chamber views. Mitral valve Doppler examination shows an increased E velocity with decreased deceleration time, and increased E/A ratio (9, 10).

The mechanism by which these two different diseases may occur at the same time remains unclear. Congenital infection including reovirus type 3 (11), genetic vulnerability to environmental precipitating factors (2), and specific HLA subtypes (12) are possible causes suggesting prenatal pathogenesis. We suggest that if the children with extrahepatic biliary atresia have a history of weakness and dyspnea, and chest radiograph films show cardiomegaly, they should be screened for cardiomyopathy.

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