

Gastroesophageal Variceal Bleeding as a Complication of Cystic Fibrosis in a 3-Month Old Patient

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Abstract- Cystic fibrosis (CF) is a hereditary disease of mucous and sweat glands, which affects the respiratory and gastrointestinal systems. Herein, we describe a 3-month-old girl with a history of recurrent episodes of urinary tract infections that required hospitalization. She was referred to our center at the age of three months, with massive gastroesophageal variceal bleeding. In physical examination, she had clubbing, hepatosplenomegaly, and mild ascites. Laboratory studies revealed high serum levels of liver enzymes and low level of Albumin. As of suspicious to CF, sweat tests were performed twice which confirmed the diagnosis of CF. Gastrointestinal bleeding due to gastroesophageal varices is a rare complication of CF, which could result as a consequence of hepatobiliary involvement of disease. Early diagnosis of CF could prevent severe complications and even death in this group of patients.

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Introduction

Cystic fibrosis (CF) is an autosomal recessive disease of mucous and sweat glands. Chronic pulmonary disease, pancreatic insufficiency, and abnormal electrolytes in the sweat are the common findings in CF patients, while infertility, delay puberty, hypoproteinemia, dehydration, and hypertrophic osteoarthropathy could be detected in some patients (1).

There is a defect in chloride ion secretion in CF patients, which is caused by mutations in the CFTR gene. The abnormal mucus could lead to a variety of clinical symptoms, including recurrent infections of airways, obstruction of the duct of the pancreas, disorder of bile secretion, and the gastrointestinal obstruction. However, CF can present with different clinical features and severity from patient to patient (1-3).

Gastroesophageal variceal bleeding is a rare, but very important complication, which could be associated with biliary atresia in CF patients. Although many patients with CF have evidence of hepatobiliary disease,

it is not associated with any clinical consequence in the vast majority of patients (1,4).

In this study, we present a 3-month old girl with CF, who was referred to our center due to massive gastroesophageal variceal bleeding.

Case Report

The patient was a 3-month-old girl, who was referred to the Children's Medical Center Hospital, the main referral pediatric center in Tehran, from a medical center in the west of Iran. She had not any problem at a birthday, but she experienced an icterus episode just 12 days after the birth, which was treated with phototherapy. Urinary tract infection was also detected at the same time. After that time, she experienced two more episodes of urinary tract infections, which required hospitalization.

At the age of three months, she was referred to our center with massive upper gastrointestinal (GI) bleeding. In physical examination, she had clubbing, hepatosplenomegaly, and mild ascites. The results of

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some laboratory studies were as follow: Prothrombin time (PT) = 16 seconds, Aspartate Aminotransferase (AST)= 150 IU/L, alanine aminotransferase (ALT)= 125 IU/L, Albumin= 2.57 gr/100ml, and Total protein= 4 gr/100ml, Hemoglobin= 5 gr/dl).

After stabilizing the patient, GI bleeding was controlled by variceal decompression. Endoscopic Variceal Ligation (EVL) was used to relieve portal hypertension, which was effective in controlling variceal hemorrhage.

Abdominal sonography was performed, which confirmed hepatomegaly, splenomegaly, and moderate ascites. Hyper echo lesion with the posterior shadow of the gallbladder was compatible with a diagnosis of cholecyst stone; small cystic lesions of the pancreas were also seen in sonography. Histopathological findings in liver biopsy indicated score 3 of fibrosis.

As of suspicious to CF, sweat tests were performed twice that confirmed diagnosis of CF. Ursodeoxycholic acid was administrated with a dosage of 20mg/kg/day, which had beneficial effects on liver biochemistry. The patient responded to this therapy; she is clinically well at the age of 1 year and has not experienced any serious complications, but she is a candidate for liver transplantation in the future.

Discussion

CF is a common hereditary disorder of ion transport, which involves multi-organs causing different symptoms. Gastrointestinal manifestation, especially hepatobiliary involvement, is one of the most important concerns in this disease (1,4).

While liver diseases usually can occur in adolescence, such manifestations are rare in infancy (5). Hepatomegaly and mild elevation of transaminases are some findings (6), which could indicate liver involvement. Inspissated secretions in the biliary system could lead to the obstruction and even progress to focal and then multilobular cirrhosis (4).

The clinical manifestations of our patient are directly associated with liver cirrhosis as one of the serious complications of CF. There are two groups of GI manifestations in CF patients: some symptoms are directly related to the basic defect in CF and others are a secondary complication of the disease or its treatment (6). Intra- and extra- hepatic defect of biliary drainage could lead to liver disease in CF (7). Hyperviscous bile which accumulates in the biliary system could cause cholangiocyte and hepatocyte damage, leading to focal

fibrosis. Such fibrosis could progress to cirrhosis over the years (5). However, our patient complicated with score 3 fibrosis very fast, just in three months. Biliary cirrhosis usually progresses in an asymptomatic process and rarely presents with icterus, ascites, and hematemesis from esophageal varices (5).

Esophageal varices could be a serious problem, with high mortality rate considering the fact that liver disease is the commonest cause of death in CF (8). EVL is a safe, efficacious treatment for control of variceal bleeding caused by portal vein hypertension (9). In our experience, variceal hemorrhage was completely eradicated with EVL and no complication was seen afterward. Ursodeoxycholic acid therapy is also effective in dissolving cholesterol gallstones and could improve biliary secretion of bile acid (10).

While GI bleeding due to gastroesophageal varices is a rare complication of CF in infancy, it should be early diagnosed and managed, while the delay in starting appropriate treatment results in severe complications and even death in this group of patients.

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