

Case report**CAROLI'S DISEASE OR SYNDROME?**

Tariq H al-Safi ,MRCP

Summary:

21 years old male patient presented with progressive jaundice for 3 months. He had H/O lieno-renal shunt for portal hypertension before 7 years . current investigations suggest the diagnosis of caroli's disease with portal hypertension. such association is analyzed & discussed in this report .

Introduction

Congenital biliary cysts are classified into five types according to Todani classification . Type I is a diffuse cystic dilatation of the common bile & hepatic ducts distal to hepatic bifurcation ,& this is the most common type . Type II shows only supra-duodenal diverticulum with normal other biliary tree. type III has isolated intra-duodenal diverticulum. & Type IV present in one of two forms : Type IV a ; have both intra & extra-hepatic cysts. & Type IV b ; have multiple extra-hepatic cysts only. Type V is called Caroli's disease in which there are multiple intra -hepatic ductal cysts only without extra-hepatic dilatation (1). if abnormalities in type V are associated with congenital hepatic fibrosis or renal cysts the disease is then called Caroli's syndrome(2).

The leading symptoms of the disease are :

Cholestasis, right upper quadrant pain & abdominal mass(3) .

Morbidity, & mortality of the biliary cystic disease correlates with type of the cystic anomaly seen, accordingly accurate diagnosis of the specific type is mandatory(4).

Manifestations of portal hypertension could also complicate the picture of caroli's syndrome because of possible association with congenital hepatic fibrosis .

In our patient, current investigations could not reveal the cause of portal hypertension , all possibilities are discussed accordingly.

Case report:

21 years old male student admitted to our GIT center on 8 April 2000 following 3 months history of progressive jaundice, associated with severe itching , arthralgia , & tea colored urine. Stool was clay in color suggestive of biliary obstruction. There was also mild abdominal pain , with intermittent episodes of fever & occasional rigors responding to antibiotics. There was no weight loss, or vomiting . the patient has also history of jaundice when he was few days old , no details were available for it's cause. Patient remained asymptomatic until 1990 (11 years old) when he started to have short episodes of malaena, continued until 1994, when he suffered progressive ascites developed over a period of 4 months . at that time diagnosis of portal hypertension was made & lieno-renal shunt surgery done. His surgery was followed by good recovery & he remained free of symptoms until his current presentation in April 2000 .

On examination at date of admission he was deeply jaundiced with scratching skin marks. The liver was enlarged (2cm below right costal margin) & firm in consistency but there was no leg edema , ascites, or stigmata of chronic liver disease.

Dr Tariq H H Al-Safi , MRCP , Gastroenterology center , Al-shaheed Adnan Hospital
Tel: + 964-1-414 1052, Fax: + 964-1-415 4642 , Email: union3@uruklink.net
Gastrointestinal & Hepatology center , Baghdad , IRAQ

Total serum bilirubin was 34mg /100ml (direct 24 mg /100ml). Liver enzymes were elevated (SGOT & SGPT, 80 & 34 units/100ml respectively). Alkaline phosphates was 35 KA units / 100ml. PT was 16 sec. & a control of 13 sec. normal serum proteins, blood picture & Bioche-mistry. immune & virological studies were normal.

Ultra-sound & Doppler studies demonstrated hepato-splenomegaly with hugely dilated portal & splenic veins & incomplete patency of the lieno-renal shunt.

MRI & MRV study confirmed complete thrombosis of portal vein & distal lieno-renal shunt. Upper Endoscopy showed grade II esophageal varices.

Liver biopsy showed large bile duct obstruction with evidence of cholestasis, but normal liver architecture.

ERCP done on 18.4.2000 reported normal pancreatic duct, but no visualization of hepatic ducts above the level of hepatic confluence, & the gall bladder was visualized with sclerosed cystic duct.

PTC done on 12.5.2000 confirmed findings of Caroli's disease with evidence of cut at level of hepatic confluence in the porta hepatis with diffuse saculation of intra-hepatic ducts.

Discussion:

1. Diagnosis of Caroli's disease is usually done radiologically, ultra-sounds & CT scans may be considered as a useful initial studies, but many patients require further study with ERCP or PTC to confirm the diagnosis(5). In this case; U/S, including Doppler study, MRI & MRV exam didn't help to confirm the diagnosis, suggesting the limited value of such investigations in this disease.

ERCP has demonstrated difficulty in visualization of the upper hepatic ducts, but although this finding was helpful in suggesting the need for further investigations, it didn't give a specific diagnosis.

PTC was more helpful since it showed the specific findings of Caroli's disease, this has indicated the superiority of PTC on other investigations in diagnosis of this disease. That may be correct because of the high level of the lesion in the biliary tree, & the difficulty of

communication of the lesion with lower biliary system imposed by pressure effect of the biliary cysts on the bile ducts.

2. Diagnosis of portal hypertension in our patient was suggested by presence of splenomegaly, & esophageal varices, and confirmed by U/S & Doppler study of the portal system. Its cause was not clear since there was no clinical evidence of chronic liver disease, as the liver biopsy showed normal liver architecture, moreover presence of portal vein thrombosis look to be secondary event since this finding hasn't been reported during previous shunt surgery in 1994, there is also no evidence of hepatic vein obstruction by his Doppler examination.
3. However association of Caroli's disease with portal hypertension need to be explained. This may necessitate repeating his liver biopsy as several reports has indicated difficulties in interpreting the histopathological material in Caroli's disease (6).
4. Looking in the literature, many incidents of frequent associations were mentioned between Caroli's disease & non cirrhotic portal hypertension without adequate explanation(7), therefore it must be concluded that future studies should concentrate in this field.

References:

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