CAROLI'S DISEASE – A CASE REPORT

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Caroli's disease, defined as a congenital dilatation and ectasia of segmental intrahepatic bile ducts in the absence of other histological abnormalities, exists in two forms: one is associated with congenital hepatic fibrosis and the other is a simple form occurring alone. The latter type is uncommon and is generally confined to a part of the liver only. Other conditions, including a choledochal cyst and renal cystic disease are frequently associated.[1, 2] A case of a young female with Caroli's disease is reported.

CASE REPORT

A 20-year-old female patient was referred to hospital with a history of recurrent episodes of intermittent fever, associated with malaise, chills and sweating for the last few years. Abdominal pain and mild icterus sometimes accompanied the pyrexia. Each of the episodes responded to conservative treatment with broad spectrum antibiotics. A cholecystectomy had been carried out for presumed acalculous cholecystitis and subsequent to surgery, the patient was well for a while. In the meantime, she got married and delivered a healthy baby by Caesarean section; the pregnancy was uneventful. There was no other significant history and the personal and family history was non-contributory. Clinical examination was unremarkable. Investigations revealed normal haematological indices and normal renal and liver biochemistry. Ultrasonography revealed hepatomegaly with multiple dilated elongated cystic structures in both lobes of liver, suggestive of dilated intrahepatic biliary radicles. Endoscopic retrograde cholangiopancreatography (ERCP) revealed saccular dilation of the intrahepatic biliary ducts involving most of the hepatic segments and without any narrowing of the common bile duct (Figures 1 and 2). Gastroscopy was normal. A diagnosis of Caroli's disease, simple type, with diffuse bile duct ectasia, was made.

DISCUSSION

Caroli's disease is an autosomal recessive disease and is more common in women. Although it is a congenital disease, it usually presents in young adults and only a few cases have been reported in children. Clinical suspicion

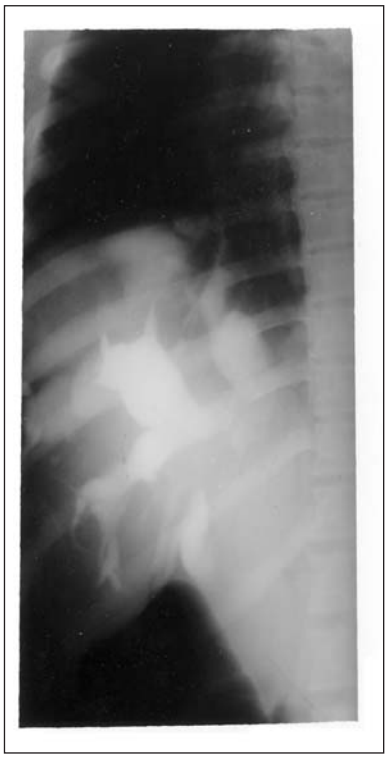

FIGURE 1
ERCP-1 showing dilated intrahepatic biliary ducts.

FIGURE 2
ERCP-2 showing the common bile duct and dilated intrahepatic biliary radicles.

of Caroli's disease is generally delayed, owing to its rarity. The major clinical feature of the disease is recurrent cholangitis. Caroli's disease is often not considered in the differential diagnosis of cholangitis, and this results in a protracted course of the disease. Intrahepatic calculi, hepatic abscess formation and secondary amyloidosis may complicate Caroli's disease. Primary cholangiocarcinoma complicates Caroli's disease in approximately 7% of cases.[1] An acute episode of cholangitis in pregnancy can be serious and potentially lethal to both the fetus and mother.[3]

The treatment depends on the clinical features and the location of the biliary abnormality. Prophylaxis of recurrent infective cholangitis is not recommended because of unpredictable results. When the disease is localised to one hepatic lobe, hemi-hepatectomy relieves the symptoms and appears to remove the risk of malignancy. In diffuse Caroli's disease, treatment options include an endoscopic sphincterotomy, hepatico-jejunostomy, and a Roux-en-Y choledochojejunostomy. These procedures help to diminish stasis and facilitate passage of biliary calculi but do not prevent the episodes of bacterial cholangitis. Liver transplantation is the best alternative when the diffuse form, without recurrent infectious cholangitis, is present.[4]

A cholecystectomy and biliary drainage give favourable results in presence of calculus disease.[5] The prognosis of the Caroli's disease is variable. Cholangitis and other clinical expression of the disease tend to resolve with treatment. However, if recurrent cholangitis is frequent in spite of medical therapy, the outlook is poor and a significant number of these patients die 5–10 years after the appearance of cholangitis.

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