Intrahepatic Biliary Dilatation with Aortoarteritis

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Intrahepatic biliary dilatation or Caroli's disease is a rare clinical entity of unknown etiology. It consists of dilatation of intrahepatic biliary ducts which communicate with normal common bile duct system. It may manifest at any age but usual age of presentation is late childhood or early adult life. The clinical features of disease includes recurrent abdominal pain associated with fever, hepatomegaly and mild jaundice secondary to recurrent cholingitis(l). It may be associated with liver disorders such as congenital hepatic fibrosis(2) and hepatic carcinoma(3). Occasionally it may be associated with renal tubular ectasia and renal cysts(4). To our knowledge it's association with aorto arteritis has not been reported.

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We present a 10 year old boy who was diagnosed to have Caroli's disease with aortoarteritis involving abdominal aorta and both renal arteries.

Case Report

A ten year old Muslim boy presented with history of not growing well for 4-5 years, recurrent high grade fever with chills and rigors associated with pain abdomen for 3 years and generalized convulsions off and on for one month. Up to the age of 5 years he was completely alright. He received various hematinics and multivitamin tonics from general practitioners for not growing well. His mother and father were first degree cousins. His two elder sibs died at age of 6 and 8 years due to some seizure disorder and the details of illness are not known. The other two sibs were completely normal at age of 6 years and 4 years. His developmental mile-stones were normal.

Physical examination revealed a poorly built but well nourished boy with normal facial features. His weight was 25 kg and height was 112 cm. His pulse rate was 110 per minute regular, good volume in all the extremities. He was hypertensive with a blood pressure of 220/150 mm of Hg in left upper limb. BP in right upper limb was 224/150 mm of Hg while BP in both lower limbs was 200/140 mm of Hg. Per abdominal examination showed a hepatomegaly of 6 cm below right costal margin in midclavicular line with firm consistency and smooth surface with a liver span of 16 cm. Spleen was one cm below costal margin. There was no clinically evident jaundice or skin lesions. Rest of the systemic examination was within normal limits.

Investigations revealed a hemoglobin of 9 g/dl with normal total and differential leucocytic count. Peripheral smear showed

microcytic hypochronic anemia. Sedimentation rate was 18 mm at end of first hour. Blood urea and serum creatinine were 190 mg/dl and 3.6 mg/dl, respectively with a glomerular filtration rate of 18 ml/minute/ 1.73 meter^2. Liver function test including S. bilirubin, SGPT, serum alkaline phosphatase were normal. Serum VDRL test was negative. Routine urine examination and urinary catecholamines were within normal limits. Among radiological investigations, ultrasound abdomen revealed bilateral contracted kidneys with smooth outline, hepatomegaly with dilated intrahepatic biliary radicles with normal common bile duct. The same findings were confirmed by abdominal CT scan. Aortogram showed very small renal arteries and nephrogram was not seen even in delayed films indicating very poorly functioning bilateral kidneys(Fig. *1).* Endoscopic retrograde cholangiography was performed under general anesthesia and revealed dilated intrahepatic bile ducts which were communicating with a normal common bile" duct *(Fig. 2).* Head CT and X-ray film of chest were normal. X-ray films of various long and short bones revealed widening and irregularity in metaphysis. Other parts of skeleton including vertebrae and skull were normal.

He was diagnosed to be suffering from Caroli's disease with aortoarteritis, chronic renal failure and started on antihypertensives in form of hydraiazine and nifedipine with supportive care for chronic renal failure. During follow up over a period of 1.5 years he required frequent hospitalizations for fever and pain abdomen and occasionally for convulsions. Along

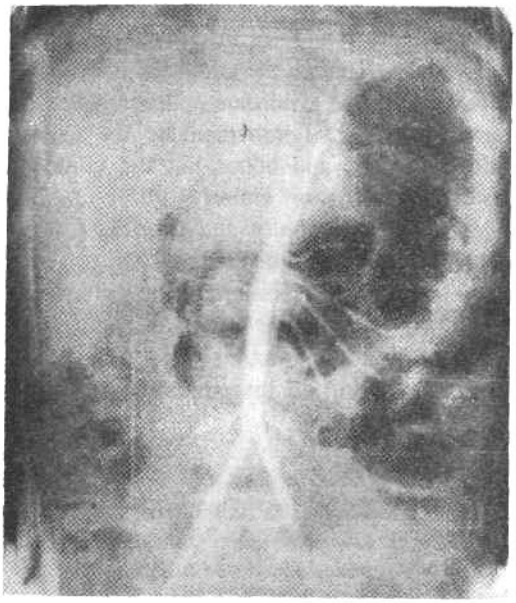

Fig. 1. Aortogram showing marked narrowing of bilateral renal arteries and abdominal aorta.

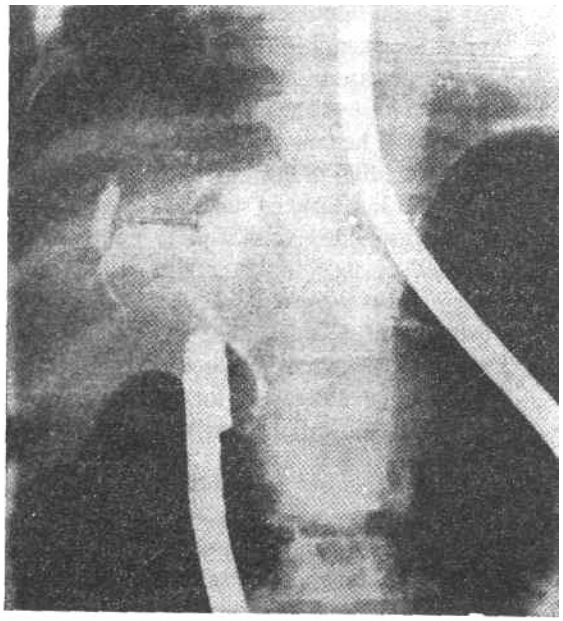

Fig. 2. ERCP showing saccular dilatation of intrahepatic biliary ducts with normal common bile duct.

with antihypertensives and antibiotics he required blood transfusion thrice. His radial pulse in left upper limb showed decrease in volume over a period of 1 year. His renal functions deteriorated progressively while liver functions remained, within normal limits. At age of 11.5 years he developed intractable seizures following stoppage of antihypertensive for 2 days and expired before he could be transferred to hospital.

Discussion

Caroli's disease is diagnosed by demonstration of dilated intrahepatic ducts with normal CBD. Ultrasonography and CT scan may demonstrate dilated intrahepatic ducts but should be confirmed by ERCP(5). Presence of abscess or stones which are complication of the disease may be seen in ultrasonography or CT scan. In our patient there were dilated intrahepatic channels with normal bile ducts. No abscess or stone could be demonstrated. The course of Caroli's disease is variable. The patient may survive for long. The cause of death most of the times is hepatic abscess or Gram negative septicemia; in few cases hepatic failure may occur in advanced stage of disease. Our patient responded very well to antibiotics for each episode of cholingitis. His liver function tests were normal till the end. The treatment of Caroli's disease consists of hepatic lobectomy in localized illness, repeated laparotomies for drainage of abscess or removal of stones(6).

The diagnosis of obstructive aortoarteritis in our patient was based on demonstration of narrowing of aorta and both renal arteries in aortagram(7). In ultrasound both the kidneys were contracted with smooth outline suggesting chronic ischemia secondary to renal arterial obstruction. The etiology of obstructive aortitis is not very clear. The suggested causes include infective, tubercular, toxic, immunologic, etc.(7,8). The course of illness depends on individual vessel involvement and the severity of illness. In our patient there was marked narrowing of renal vessels at the time when he presented to us. He expired due to end stage renal disease secondary to severe ischemia. The treatment for aortoarteritis is not well defined and includes antinflammatory agents, antitubercular drugs and surgical intervention.

Explanation for occurrence of two rare but well defined clinical conditions of unknown etiology in our patient is difficult. An autosomal dominantly inherited syndrome involving intrahepatic biliary ducts with vessels and bones has been reported by Alagille *et al.(9).* This syndrome consist of intrahepatic biliary atresia with vascular involvement in form of pulmonary stenosis peculiar facies and vertebral anomalies. This usually manifests in early infancy as cholestasis. Though the anatomic structures involved in this syndrome and our patients are similar, *i.e.,* intrahepatic biliary ducts, blood vessels, and bones but the clinical manifestations are entirely different. In view of consanguinity and deaths of two sibs the combined disease in our patient may be regarded as variant of the syndrome described by Alagille *et al.* However, a possibility of some toxic or immunological factor involving all the structures together cannot be ruled out.

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Klippel Trenaunay Weber Syndrome Associated with Abdominal Hamartoma with Undescended Testis

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Klippel Trenaunay Weber Syndrome is a non-heritable disorder consisting of a macular vascular nevus (port-wine nevus) in combination with bony and soft tissue hypertrophy and venous varicosities(l). Though a few cases have been reported(2-4), but to the best of our knowledge, the association of an abdominal hamartoma with undescended testis and appendage in this disorder has not been documented.

Case Report

A 2!/2-year-old boy born of non-consanguinous parents was admitted to our hospital in October, 1991 with history of progressive enlargement of left lower limb, buttocks, perineum; pink stains at different sites of the left side of the body since birth

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