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A CASE OF CAROLI'S DISEASE

General Medicine

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ABSTRACT

Caroli's disease is a rare inherited disorder. It has two types. The first is characterized by dilatation of the bile ducts, while the second form is associated with hepatic fibrosis and portal hypertension and is known as Caroli's syndrome. Patients with these syndromes may have recurrent episodes of cholangitis and are also at risk for associated bacteremia, sepsis and cholangiocarcinoma. However, no specific treatment is available as yet. We present a case of an 18 year old male patient who presented with complaints of right sided abdominal pain and was diagnosed as Caroli's disease to make a point that there is need to develop some guidelines on management of these cases to improve their quality of life and regress progression to malignancy as although rarely seen, this entity has potential to badly affect quality of life of the patients and turn malignant.

KEYWORDS

Caroli's disease, Intrahepatic biliary radicles

INTRODUCTION:

Caroli's disease is a rare disorder described by dilatation of the intrahepatic biliary ducts. Jacques caroli, a gastroenterologist, initially depicted an uncommon inherent condition in 1958 in Paris, France, as "nonobstructive saccular or fusiform multi-focal segmental dilatation of the intrahepatic bile ducts. Caroli's disease is also known as communicating cavernous ectasia or congenital cystic dilatation of the intrahepatic biliary tree. There are two forms of Caroli's diseases, the most well-known being the isolated situation where the bile ducts are extended by ectasia, which is also called as type 1 CD. While the second form is generally known as Caroli's syndrome or type 2 CD, which is characterized by presence of portal hypertension and fibrosis in the liver, also commonly known as congenital hepatic fibrosis. Hepatic failure and polycystic kidney disease are also reported to be associated with it. It has even been regarded as precancerous and patients with type 2 CD are reported to have a 2.5–17.5% likelihood of developing cholangiocarcinoma,[1] which is 100-fold higher than in subjects with normal hepatobiliary ducts and 10-fold higher than in those with calculi.[2] We present this case of Caroli's disease as it has significant morbidity associated and needs attention from medical community to give specific guidelines for its management to regress progression to complications.

CASE:

An 18 year old male was admitted in male medicine ward with complaints of pain in right hypochondrium and right iliac fossa radiating to umbilicus since four days. There was no history of fever, nausea, burning micturition, cough or diarrhoea. There was no significant past history. He belonged to lower socio economic class and was a labour by occupation. He had a habit of occasional gutkha chewing but was non-smoker and also not an alcohol consumer. Both his parents were labourer and both of them were reported to have positive human immunodeficiency virus status. Both of them had succumbed to tuberculosis a few years back.

On examination, the patient was well built and well nourished. He was afebrile. Pulse was 82/minute, regular with normal force and volume. His blood pressure was 100/60 mm/hg in right arm in supine position. He had pallor but there was no icterus, cyanosis, clubbing or lymphadenopathy. Respiratory cardiovascular and central nervous system examination was normal. On per abdomen examination there was tenderness present over the right hypochondrium and right iliac fossa. However, there was no organomegaly. Lab investigations revealed mild anaemia and elevated alkaline phosphatase. USG abdomen and CT Abdomen showed changes of Caroli's disease.

TABLE - 1

Investigations(23/6/18)	Values
Hb	10.1 mg/dl
Total leucocyte count	7300/cu.mm
Platelets	2.23 lacs/cu.mm
Serum iron	62 mcg/dl
Serum ferritin	28 mg/dl
Total iron binding capacity	358 mcg/dl
Reticulocyte count	0.5%
Blood urea	41 mg%
Serum creatinine	0.9 mg%
Serum sodium	134 mmol/l
Serum potassium	4.3 mmol/l
Serum chloride	104 mmol/l
Urine – routine & micro	Albumin – trace Sugar – absent Pus cells- 2-3 Epithelial cells- 2-3
Total	0.5 mg%
Bilirubin	
Direct bilirubin	0.2 mg%
Indirect bilirubin	0.3 mg%
SGPT	16 iu/l
SGOT	24 iu/l
Serum alkaline phosphatase	195 u/l
HIV	Negative
HCV	Negative
HBsAg	Negative

Peripheral smear	(23/6/18)
RBC	Predominantly normocytic normochromic, occasional target cells seen
WBC	Normal
Platelets	Adequate on the smear

Table - 2

Chest x-ray	Both lung fields appear normal, , both domes of diaphragm and costophrenic angles are clear, heart size appears normal, bony thorax appears normal
Ultrasonography of abdomen and pelvis	Dilated gall bladder and dilated intrahepatic biliary radicles filled with sludge represents infective aetiology. Callus noted in intrahepatic biliary radicles and common bile duct. Raised cortical ecogenecity in right kidney represents inflammatory changes. Urinary bladder shows changes of cystitis, very minimal intrahepatic free fluid
CT abdomen and pelvis	Multiple cystic dilatation of IHBR with small portal radicles passing within, suggestive of choledocal cyst; possibility of caroli's disease Few ca lculi seen within cystic dilated IHBR , few calculus seen in common biliary ducts and hepatic ducts, cystitis (figure 1,2)

He was treated with ursodeoxychoic acid and symptomatic and supportive treatment like anti spasmodics, till the stay in hospital. His symptoms improved and was discharged with advice of regular follow up.

DISCUSSION

Caroli's disease influences around 1 in 1,00,000 individuals, but Caroli's syndrome is relatively more commonly seen. In Caroli's disease, polycystic segmentation of the larger intrahepatic bile ducts (segmental, left and right) occurs, whereas in Caroli's syndrome the ductal abnormalities are more widespread. Both forms are characterized by fibrosis of the portal tracts and are thought to originate from a pathologic development in the formation of the ductal plate. The ductal plate originates from hepatocytes surrounding the intrahepatic portal vein branches. Further differentiation during fetal life results in the formation of small tubules, which coalesce to form the intrahepatic biliary tree. Failure of the ductal plate to differentiate results in ductal plate malformations. The anomalous structure of the intrahepatic bile ducts leads to saccular or fusiform dilatation and ectasia with biliary stasis, cholelithiasis, cholangitis with intrahepatic abscesses and recurrent sepsis. Although the underlying cause is poorly understood, they are thought to be genetic conditions. Caroli's disease generally occurs sporadically in people with no family history of the condition; however, rare reports of autosomal dominant inheritance in association with autosomal dominant polycystic kidney disease do exist.

People affected by this condition experience recurrent episodes of cholestasis and bacterial cholangitis. They may also have hepatic fibrosis and portal hypertension[3,4,5].

The first symptoms typically include fever, intermittent abdominal pain which can be radiating to adjacent areas of the abdomen and recurrent jaundice. Caroli's disease has been reported to be associated with autosomal recessive polycystic kidney disease (ARPKD), cholangitis, gallstones, biliary abscess, septicemia, liver cirrhosis, renal failure and cholangiocarcinoma. It is also associated with 100 times more risk for cholangiocarcinoma than the general population. Portal hypertension may be present, resulting in splenomegaly, hematemesis and melena. Although surgery may be indicated in some cases, the progression to complications like recurrent calculi and cholangitis can be regressed with medication like ursodeoxycholic acid. Regular follow up is needed to screen for other complications.

In short, Caroli's disease is a congenital disease which may significantly affect the quality of life of the patient and has high propensity to progress to malignancy. Early diagnosis of this disease and treatment may regress formation of calculi and progression to other complications improving the quality of life of these patients. Studies are required to introduce some more modalities of treatment for this disease to reduce the associated morbidity.

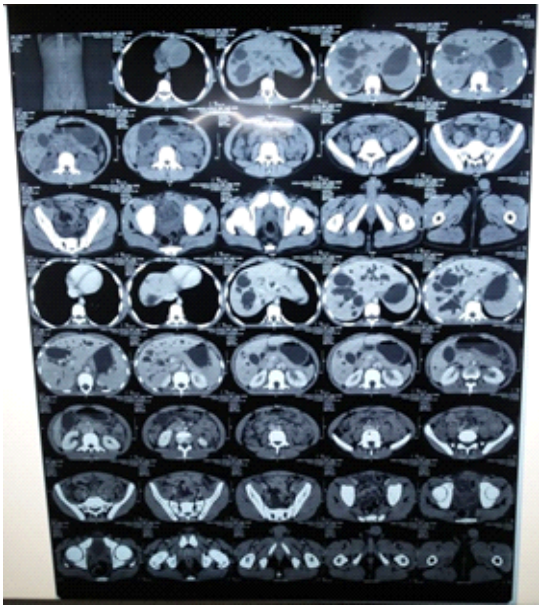

FIGURE 1 (26/6/18)

FIGURE 2 (26/6/18)

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