Multicystic Hepatic Lesion: An Unusual Presentation of Extra-Pulmonary Tuberculosis in a Child

Tuberculosis of the liver is a rare form of extra pulmonary tuber-culosis, and is seen more commonly in immunocompromised patients or in association with disseminated tuberculosis. Liver involvement without involvement of lung or other organs is rare. Nodular form of liver tuberculosis presenting as abscess is uncommon, and is commonly mistaken for pyogenic/amebic liver abscess or malignancy. Definitive diagnosis requires detection of tubercular bacillus in pus or liver biopsy [1].

A 12-year-old girl presented with non-localized upper abdominal pain for 3 months, with history of fever off-and-on and weight loss of 4 kg in two months. There was no history of previous hospitalization or contact with tuberculosis. Immunization was complete as per national immunization schedule; however, BCG scar was absent. On examination, child was stunted and wasted (weight for age at –2.04 *z*-score, height for age at –2.54 *z*-score as per IAP charts). General physical examination revealed severe pallor, angular cheilitis, mucositis and knuckle hyperpigmentation, with no lymphadenopathy. On systemic examination, there was hepatomegaly with other systems being unremarkable. Chest radiograph was normal. Ultrasonography abdomen revealed a large heterogenous solid cystic mass lesion involving the segment VIII and IV of liver, extending till the subcapsular regions. A possibility of hydatid cysts, multiple pyogenic abscesses and fungal abscesses was kept. On laboratory evaluation, hemogram was performed: Hemoglobin 4.4 gm/dL, total leucocyte count 17200/mm^3 (lymphocytes 18%, neutrophils 78%) and peripheral smear revealed dimorphic blood picture with microcytic hypochromic and macrocytic normochromic red cells. ESR was raised (60 mm/h); liver and renal function tests were normal. Serum iron levels (40 mcg/dL) and serum B12 levels (160 pg/mL) were both low. Mantoux test and gastric aspirate for cartridge based nucleic acid amplification test (CBNAAT) were negative. Stool microscopy did not reveal cyst or ova and HIV test and immunodeficiency work up was negative. Computed tomography (CT) scan of abdomen revealed a large ill-defined heterogeneously hypodense mass lesion involving the left and right lobes (segment VIII and IV) of liver, faintly hyperdense internal septations could be seen (**Fig. 1**). A possibility of hydatid cysts and malignancy was kept. Hydatid serology was negative. CT-guided tru-cut needle biopsy was planned, for which the child was referred to a higher centre. Liver biopsy showed multiple epithelioid cell granulomas, positive for acid-fast bacilli on Ziehl Neelson (ZN) stain. A diagnosis of tuberculosis was made and child was started on directly observed treatment, short-course (DOTS) therapy for tuberculosis. On follow up after 2 months, child started gaining weight and repeat ultrasonography showed decreasing size of liver abscesses.

Primary hepatic tuberculosis without pulmonary or miliary tuberculosis is an uncommon diagnosis. The diagnosis is frequently missed due to lack of suspicion and can mimic other etiologies like bacterial, amebic or fungal liver abscess [2]. In a study from South Africa, in 296 patients with hepatic tuberculosis, tubercular abscess accounted for only 0.54% cases [3]. In an Indian study of 242 immunocompetent tuberculosis patients, 38 had liver involvement, of which 10 had tubercular liver abscess [4]. Patients usually present with fever, abdominal pain, anorexia, hepatomegaly and loss of weight with jaundice being an uncommon presentation. Right lobe of liver has been found to be more commonly involved (82.5%) [1].

Radiological findings are variable and insufficient for diagnosis. Majority of the cases have shown heterogenous, anechoic or hypoechoic lesions with irregular margins; however, some reports have described a hyperechoic mass [5]. Amebic or pyogenic liver abscess or hepatocellular carcinoma are the differential diagnosis. Definitive diagnosis can be made by detection of tubercular bacilli in pus or liver biopsy stained by ZN stain [1]. Although culture is the gold standard, but it requires long incubation period. Polymerase chain reaction has a sensitivity of 92.4% and specificity of 98%, and should be performed for rapid diagnosis [6].

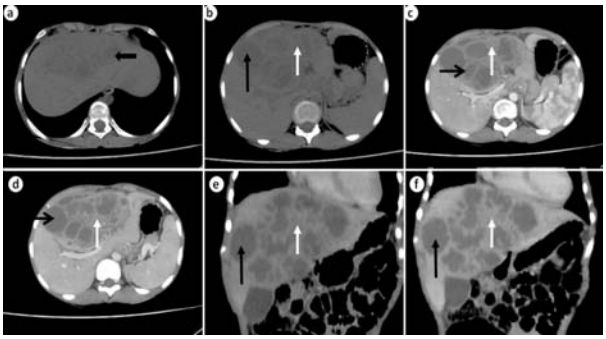

Fig. 1 (*a*) Axial plain CT images reveal a large ill-defined heterogeneously hypodense mass lesion involving left and right lobes (segment VIII and IV) of liver; (*b*) Cystic appearing areas (black arrow) as well as solid slightly hyperdense areas (white arrow) seen within the lesion. Contrast enhanced CT scan shows axial images: late arterial phase (*c*) and porto venous phase (*d*), composed of solid and cystic areas. Cystic areas (black arrow) show no significant enhancement whereas solid areas show mild progressive enhancement (white arrow); Contrast enhanced CT scan shows coronal images of the lesion. (*e*) late arterial phase, (*f*) porto venous phase. Lesion is composed of solid and cystic areas. The cystic areas (black arrow) show no significant enhancement whereas solid areas show mild progressive enhancement (white arrow).

We report this case to highlight a rare manifestation of a common disease. A high index of suspicion may help in timely diagnosis and avoid unnecessary investigations or surgical intervention.

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Benign Recurrent Intrahepatic Cholestasis - Unravelleing the Paradox

Benign recurrent intrahepatic cholestasis (BRIC) is a rare autosomal recessive bile salt transport disorder, characterized by recurrent episodes of pruritus, cholestatic jaundice with normal or low gamma glutamyl transpeptidase (ggt). Bric and progressive familial intrahepatic cholestasis (PFIC) represent two extremes of a continuous spectrum of genetic intrahepatic cholestatic disorders. The exact prevalence of BRIC still remains unknown. BRIC can present at any age but usually before the second decade [1] with disturbing pruritus as the primary symptom. Very few case reports have been published in Indian literature [2,3]. We report the clinico-laboratory profile and follow up of seven patients with BRIC seen over a period 20 years (2000-2020).

Patient 1: A 16 year old adolescent boy, 1st born to 3^0 consanguineous parents, presented with 2 weeks history of severe pruritus associated with mild jaundice. His first cousin died of cholestatic liver disease at the age of 7 years. On examination, he was well nourished with icterus and scratch marks on his skin. There was no hepatosplenomegaly. Complete hemogram and renal profile were normal. His total serum bilirubin was 5.1 (direct:4.1) mg/dL, alamine aminotransferase (ALT) was 133U/L, aspartate aminotransferase (AST) was 143U/L, γ-glutamyl transferase-20 U/L, serum alkaline phosphatase-268 U/L, cholesterol -145 mg/dL and serum bile acids 491 µmol/L (0.5-10). Prothrombin time was normal. USG abdomen showed normal liver echogenecity and intrahepatic radicles were not dilated. Liver biopsy was deferred due to refusal of consent. In view of the strong positive family history of cholestatic liver disease, gene testing was done that showed a single heterozygous missense mutation [c.1244A>G] in exon 13 of *ATP8B1* gene confirming BRIC type 1. He was treated with rifampicin for 3 weeks, and at 5 months follow up, all his laboratory parameters were normal.

Patient 2: A 16-year-old boy presented with intense pruritus and jaundice for 20 days. There was history of two stereotypical episodes of pruritus associated with cholestatic jaundice in the last 1 year. During the icterus free intervals, he was normal. He was second born to second degree consanguineous parents, and his elder sister had been diagnosed as BRIC type 1. On examination, he was well nourished with deep icterus and scratch marks on his skin. There was no hepatosplenomegaly. Investigations revealed normal hemogram and renal profile. Total bilirubin was 31 (direct: 25.3) mg/dL. ALT, AST and total protein were normal. Serum bile acids were 350 µmol/L and GGT was 12 U/L. Magnetic resonance cholangiopancreatography (MRCP) showed mild hepatomegaly without intra or extra hepatic biliary dilatation. Liver biopsy showed marked canalicular cholestasis, mild lobular inflammation, with intact interlobular bile ducts and no fibrosis. He was treated with ursodexycholic acid (UDCA) and rifampicin, and at 4 weeks his jaundice had cleared. Genetic testing showed a homozygous missense mutation [c.922G>A] in exon 10 of *ATP8B1* gene confirming BRIC type 1. He is under follow up for the last three years, and is doing well without any worsening.

In the remaining five patients (3 girls) with BRIC diagnosed histologically, the median age at onset of symptoms was 11 (range: 8-18) years. The cholestatic episodes varied with an average of 1 to 3 per year and the reported asymptomatic periods were lasting for a maximum period of 3 years. There was history of consanguinity in 80%, of which second and third degree consanguinity was seen in 50% each. One child was adopted. Liver histology done in all five patients showed intrahepatic cholestasis with intact interlobular bileducts and no fibrosis. All were treated with UDCA, rifampicin and cholestyramine in varying combinations. Over these 20 years, 2 girls got married and both had pruritus during pregnancy. Another boy diagnosed with BRIC at 8 years was given complementary and alternative medicine for refractory pruritus at 17 years, that worsened his liver function following which, the jaundice deepened, bilirubin rose to 40 mg/dL and his INR reached 4. He underwent 3 cycles of plasmapheresis, but succumbed to the illness prior to liver transplant.