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References

1. Giefer MJ, Murray KF, Colletti RB. Pathophysiology, diagnosis, and management of pediatric ascites. *J Pediatr Gastroenterol Nutr*. 2011;**52**:503–13.
2. Redman JF, Seibert JJ, Arnold W. Urinary ascites in children owing to extravasation of urine from the bladder. *J Urol*. 1979;**122**:409–11.
3. Vasdev N, Coulthard MG, De la hunt MN, Starzyk B, Ognjanovic M, Willetts IE. Neonatal urinary ascites secondary to urinary bladder rupture. *J Pediatr Urol*. 2009;**5**:100–4.
4. Kuwata T, Matsubara S, Nakamura S, Nakai H. Urinary ascites in a fetus with posterior urethral valve: antenatal diagnosis. *Pediatr Int*. 2011;**53**:281–2.
5. Murphy D, Simmons M, Guiney EJ. Neonatal urinary ascites in the absence of urinary tract obstruction. *J Pediatr Surg*. 1978;**13**:529–31.
6. Kato A, Yoshida K, Tsuru N, Ushiyama T, Suzuki K, Ozono S, et al. Spontaneous rupture of the urinary bladder presenting as oliguric acute renal failure. *Intern Med*. 2006;**45**:815–8.
7. Saliba W, Grant ME. Conservative management of spontaneous bladder rupture. *Kansas J Med*. 2011;**4**:44–6.
8. Stebbing J, Ezra DG, Cackett PD, Greenstein AS. Ascites and apparent renal failure treated with a Foley catheter. *J R Soc Med*. 1999;**92**:582–3.

Congenital hepatic fibrosis with extra-hepatic porto-systemic shunt and hepatopulmonary syndrome successfully managed with living related liver transplantation

Introduction

Congenital hepatic fibrosis (CHF) is a rare condition in which portal hypertension may present without overt signs or symptoms of liver disease. Association of CHF with hepatopulmonary syndrome (HPS) has been well described in literature,[1] however there are no reports of CHF with extra-hepatic porto-systemic shunt presenting with HPS. We report here such a patient of CHF with extra-hepatic porto-systemic shunt presenting with HPS, who was successfully managed with liver transplantation.

Case report

A 14-year-old boy presented to us with two years history of exertional dyspnea and easy fatigability. He also gave history of bluish discoloration of hands and lips for the last three months. There was no history of jaundice, ascites, gastrointestinal bleed or easy bruisability. Clinical examination revealed cyanosis, grade II clubbing and hepatomegaly. There was no jaundice and the remaining systemic examination was normal. Investigations revealed a hemoglobin of 11.8 gm%, a platelet count of 146,000/cu mm and a normal coagulogram. Liver and kidney function tests were normal except for hypoalbuminemia. Arterial blood gas analysis showed an oxygen saturation of 68% and a PaO_2 of 37 mmHg. The patient

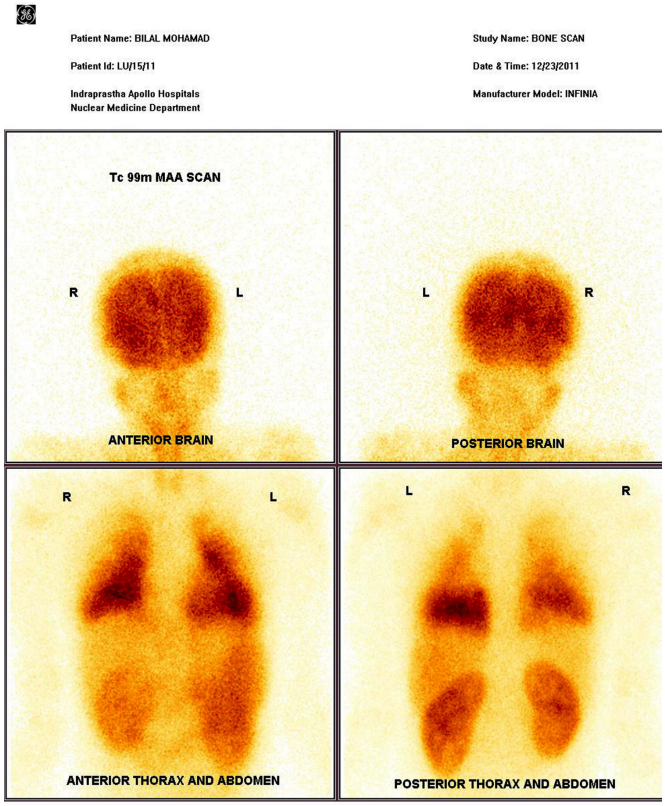

Figure 1: ^{99}Tc MAA scan showing patchy perfusion in both lungs with reduced perfusion especially in apices of both lung fields. Significant uptake (73%) in the brain is visible and suggests intrapulmonary shunting.

underwent echocardiography to rule out congenital heart disease, which showed normal cardiac chambers, ejection fraction and pulmonary artery systolic pressure. However bubble contrast echocardiography was strongly positive after four beats, suggestive of HPS. Subsequently the patient underwent a macro-aggregate albumin scan which showed a shunt fraction of 73% suggestive of severe HPS (**Figure 1**). Conventional pulmonary angiography did not reveal any major shunts amenable to embolization. Ultrasound abdomen showed heterogeneous liver parenchyma with a small caliber main portal vein. CT angiography showed heterogeneous attenuation of liver, a small caliber portal vein and a large porto-caval shunt extending from the origin of portal vein to the intrahepatic IVC, measuring 15 mm, and accompanied by mild splenomegaly (**Figures 2 & 3**). Upper gastrointestinal endoscopy did not reveal any esophageal or gastric varices. Etiological work-up for liver disease including viral and autoimmune markers, iron studies, ceruloplasmin and alpha-1-antitrypsin were all found normal. Thus a liver biopsy was done which revealed features of congenital hepatic fibrosis (**Figure 4**). The child was evaluated for liver transplantation and the mother became the prospective donor. A standard right lobe graft liver transplantation was performed and the grafted right portal vein was anastomosed with the main portal vein of the recipient along with ligation of the major shunt. The patient needed prolonged postoperative ventilatory support and gradual improvement in oxygenation with PaO_2 rising from 37 to 60 mmHg was observed by the end of two weeks. The patient was

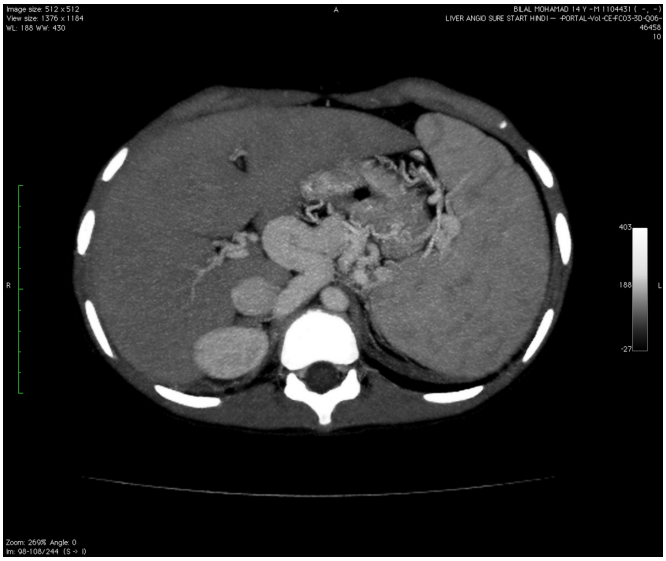

Figure 2: Maximum intensity axial projection image on contrast–enhanced CT scan showing a large porto-caval shunt extending from the origin of portal vein to the intrahepatic IVC, measuring 15 mm.

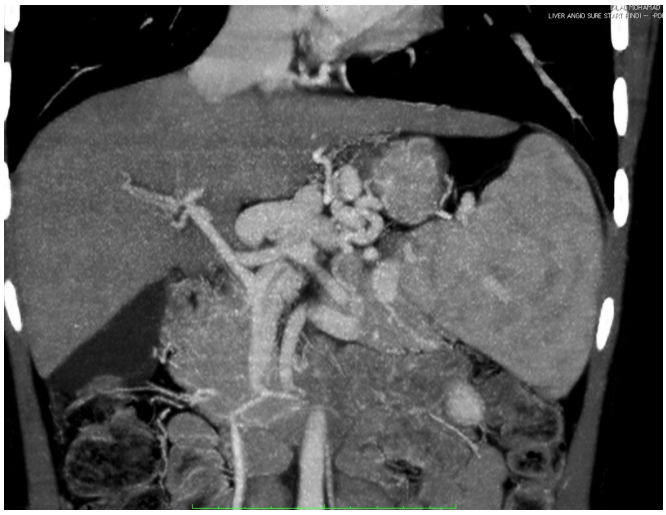

Figure 3: Maximum intensity coronal projection image on contrast–enhanced CT scan showing the small caliber of the main portal vein and absence of intrahepatic portal vein.

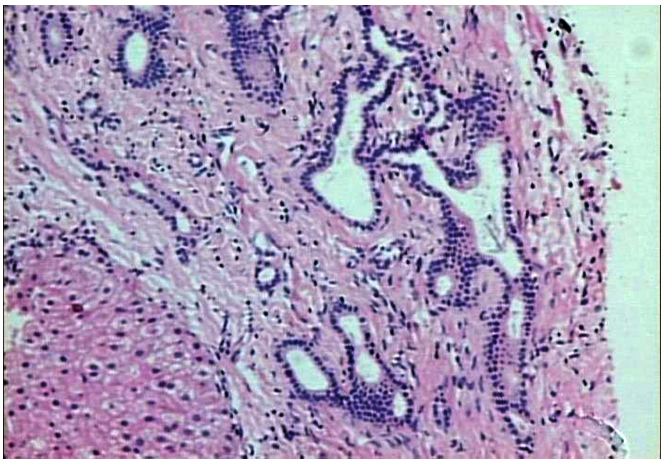

Figure 4: Liver biopsy shows wide areas of fibrosis with bile ducts. These bile ducts are angulated and dilated. Bile duct proliferation is seen at the periphery of the liver lobule along with mixed inflammatory infiltrate.

discharged on domiciliary oxygen therapy for four weeks. At follow-up after five months, the child is maintaining normal oxygenation at ambient oxygen levels and is back to his regular activities.

Discussion

There are very few reports on the association of congenital hepatic fibrosis with hepatopulmonary syndrome and extra-hepatic porto-systemic shunts. Currently liver transplantation is the only successful treatment for HPS, since no effective medical therapy is available.[2] However, postoperative mortality is high and resolution of arterial hypoxemia is often delayed in patients with severe HPS.[3] These patients usually have a longer

hospital stay and may take months to recover from hypoxemia.[4] The strongest predictors of death include a preoperative partial pressure of oxygen of 50 mmHg or less and a lung scan with brain uptake of 20% or more.[5] Our patient had a favorable outcome despite suffering from severe HPS and a significant brain uptake of 73%. Such association of CHF with extra-hepatic porto-systemic shunt and HPS precludes shunt ligation alone as a therapeutic option. Liver transplantation itself is technically demanding given the small caliber of the portal vein making portal vein anastomosis difficult. In our patient the anastomosis was successful and a good inflow was obtained after shunt ligation. The patient had an uneventful surgery but required domiciliary oxygen therapy for one month before complete recovery.

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References

1. Nagral A, Nabi F, Humar A, Nagral S, Doctor N, Khubchandani SR, et al. Reversal of severe hepato-pulmonary syndrome in congenital hepatic fibrosis after living-related liver transplantation. *Indian J Gastroenterol*. 2007;**26**:88–9.
2. Rodriguez-Roisin R, Krowka MJ. Is severe arterial hypoxaemia due to hepatic disease an indication for liver transplantation? A new therapeutic approach. *Eur Respir J*. 1994;**7**:839–42.
3. Taille C, Cadranel J, Bellocq A, Thabut G, Soubrane O, Durand F, et al. Liver transplantation for hepatopulmonary syndrome: a ten-year experience in Paris, France. *Transplantation*. 2003;**75**:1482–9; discussion 46–7.
4. Philit F, Wiesendanger T, Gille D, Boillot O, Cordier JF. Late resolution of hepatopulmonary syndrome after liver transplantation. *Respiration*. 1997;**64**:173–5.
5. Arguedas MR, Abrams GA, Krowka MJ, Fallon MB. Prospective evaluation of outcomes and predictors of mortality in patients with hepatopulmonary syndrome undergoing liver transplantation. *Hepatology*. 2003;**37**:192–7.

Appendagitis: a benign differential diagnosis in acute abdomen

Introduction

Epiploic appendages are visceral peritoneal out-pouchings of colon containing fat and blood vessels. Appendagitis is a benign and self-limiting condition of epiploic appendages.[1] Primary epiploic appendagitis (PEA) results from spontaneous thrombosis of the veins draining the appendages in absence of any torsion or ischemia.[2] Secondary epiploic appendagitis, on the other hand, develops due to inflammation of adjacent structures like appendicitis, diverticulitis or cholecystitis. PEA often presents with acute abdominal pain and can mimic acute diverticulitis or appendicitis on clinical examination. Its diagnosis is challenging due to the lack of any pathognomonic clinical features. Computed tomography (CT) scan has an important role in the diagnosis of PEA.[3] Timely diagnosis can avoid unnecessary surgical treatment.

Case report

A 35-year-old woman presented with sudden onset, severe pain abdomen since five days; localized to the upper left quadrant of the abdomen. The pain was accompanied by fever for initial two days. There was no history of constipation or vomiting. Physical examination revealed marked tenderness over the left upper quadrant of the abdomen with peritoneal sign of guarding and rebound tenderness. Rest of the physical examination was normal. All laboratory tests were within normal limits including white cell count, serum amylase, and liver and renal function tests. Pregnancy test was also negative. Ultrasound examination of the abdomen revealed a hyperechoic non-compressible pericolonic mass, and no organomegaly was seen. The persistent pain with localized peritoneal sign, prompted us to perform a contrast-enhanced CT scan of the abdomen and pelvis. The CT scan revealed focal fat stranding with mesenteric fat inflammation adjacent to the colon in the left para-sagittal, paraumbilical, omental region, which were consistent with the diagnosis of epiploic appendagitis (**Figures 1A & 1B**). Colonoscopic examination was normal. The patient was treated conservatively and she improved within a few days.