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Endotipsitis caused by extremely drug-resistant *Klebsiella pneumoniae*

Introduction

Transjugular intrahepatic portosystemic shunt (TIPS) is commonly used for the decompression of portal hypertension. TIPS infection, also known as 'endotipsitis', is a rare but serious complication of TIPS insertion.[1] To date approximately 42 cases of 'endotipsitis' have been described. To the best of our knowledge this is the first case report of endotipsitis caused by an extremely drug-resistant Klebsiella pneumoniae (XDR).[2]

Case report

A 28-year-old man with decompensated chronic alcohol-related liver disease was admitted with upper gastrointestinal bleeding. He had no history of hospitalization in the previous 6 months. He underwent TIPS insertion due to variceal bleeding which did not respond to endoscopic variceal ligation. Three days post TIPS procedure he developed ARDS with leukocytosis, hyperbilirubinemia, elevated aminotransferases, hypoalbuminemia, coagulopathy and septic shock. Two sets of blood cultures grew extremely drug resistant (XDR) *Klebsiella pneumoniae* (resistant to all antibiotics tested including carbapenems, colistin, and tigecycline). Workup, including urine culture, transthoracic echocardiography, chest radiography, abdominal imaging, and diagnostic paracentesis, failed to localize the source of the bacteremia. He initially improved with a combination of multiple antibiotics (imipenem, colistin, tigecycline, chloramphenicol, amikacin, ciprofloxacin and fosfomycin). However he had re-bleeding: Doppler ultrasound did not show any thrombus and CT angiogram revealed left gastric artery bleeding for which embolisation was performed. His TIPS was re-examined via transjugular route to look for any thrombus. Stent patency was confirmed; however in view of the recurrent bleed, TIPS stent dilatation with balloon was done. After transient improvement he again developed fever, worsening of sepsis and persistent XDR

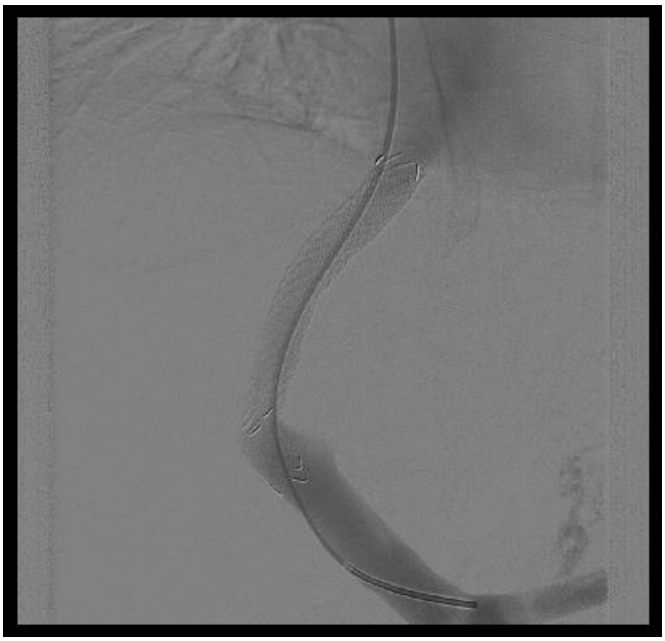

Figure 1: TIPS seen following contrast injection into the portal vein during the interventional proceedure (TIPSogram)

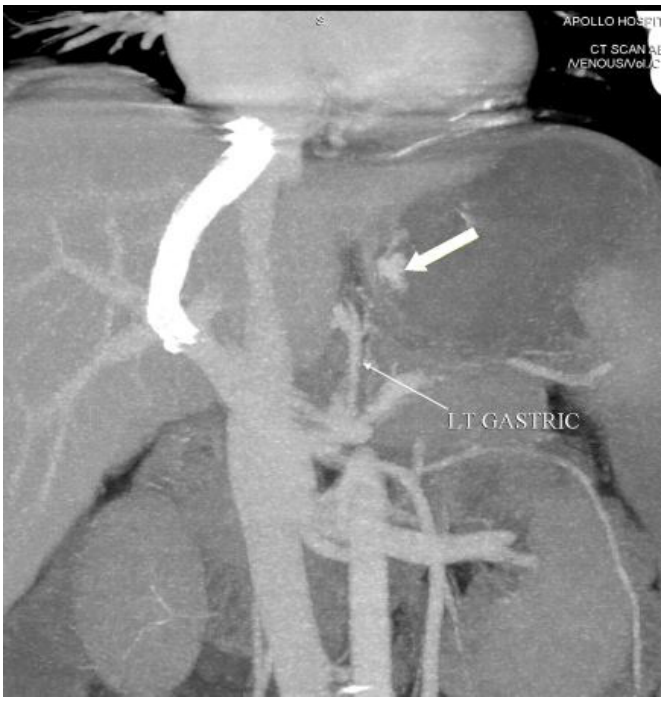

Figure 2: Coronal CECT showing contrast extravasation from the left gastric vessel TIPS in place

Klebsiella bacteremia with the same antibiotic-resistance pattern, seven days after the initial blood culture positivity without any other source of infection. Endotipsitis was diagnosed based on the time frame of infection, presence of the TIPS serving as a potential nidus of infection in the absence of other source of infection. Liver transplantation was planned but before a donor was identified, he deteriorated and expired despite multiple antibiotic courses and supportive care.

Discussion

Endotipsitis i.e. persistent TIPS infection, first described by Sanyal and Reddy in 1998[1] has an overall incidence that varies from 1.7 - 5.1%.[3] Endotipsitis can occur as an early infection if occurring within 120 days of TIPS procedure or late infection if occurring after this period. These differ in terms of the causative agents and pathogenesis.[3] Although the clinical presentation of endotipsitis is variable, fever is a constant feature. Worsening jaundice, increasing ascites, recurrent variceal bleeding and shock are also described and attributed to TIPS occlusion. Laboratory findings include leukocytosis, anemia, hyperbilirubinemia, elevated aminotransferases, hypoalbuminemia and coagulopathy similar to our case.

The exact diagnostic criteria of endotipsitis are still being debated.[4] The "gold standard" for diagnosis of tipsitis is removal of stent and culturing the stent which is possible only during liver explants at time of transplantation or autopsy. Tissue cultures obtained from direct endothelial biopsy performed from within the TIPS[5] is not widely available and is not required for the definitive diagnosis of endotipsitis. "Definite infection" as described by Sanyal & Reddy is defined as clinically significant continuous bacteremia (fever and multiple positive blood cultures) along with vegetation or thrombi inside the TIPS. "Probable infection", as in our patient, is defined as sustained bacteremia and unremitting fever in a patient with apparently normal TIPS without any other identifiable source of infection [1]. De Simone et al [6] described stricter criteria to define sustained bacteremia as two blood cultures drawn 4-12 h apart that were positive for the same organism without another identifiable source of bacteremia. Later Armstrong and MacLeod[3] put forth a more specific standardized definition requiring sustained bacteremia in a patient with a TIPS device, with or without thrombus, plus no other identifiable infective focus, after an exhaustive workup. They defined sustained bacteremia as two or more blood cultures positive for the same organism, the first and last being separated by 7 days or more. Our case fulfils the above definitions.

TIPS stenosis or thrombosis, a strong risk factor is reported in 47% cases as demonstrated by ultrasound, doppler-imaging or other methods [4]. In our case doppler ultrasound and TIPS revision was done to assess TIPS patency or TIPS-biliary fistula. Other tests included negative urine, tracheal secretions, and ascitic fluid cultures. Chest radiographs, CT abdomen and angiogram, transthoracic echocardiography, upper and lower gastro-intestinal endoscopy were done to rule out deep-seated infections.

Removal of shunt continues to be the "gold standard" treatment of endotipsitis; however, treatment is essentially medical therapy as per culture, because TIPS is irremovable without liver transplantation. In the literature, medical treatment was ineffective in the eradication of infection in 10 cases, all of who died (seven with definite endotipsitis and three with probable endotipsitis).[4]

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Giant choledochal cyst in infancy- A rare entity

Introduction

Dilatation of the biliary tree to variable extent and severity is called choledochal cyst (CDC). The prevalence is higher in Asian countries, especially Japan, and females are more affected than males in a ratio of 4:1[1,2]. Eighty percent of CDC patients manifest clinically in childhood and are either detected on an antenatal scan or present later with jaundice and/or abdominal pain. The triad of pain, lump and jaundice is rare in children. Giant choledochal cysts (GCDC), defined as >10 cm in diameter, are uncommonly encountered in practice. The reports of GCDC that are described in the literature pertain to older children and adults. This report highlights that a GCDC can present even in infancy and without features of cholestasis.

Case report

A 5-month-old boy presented with failure to thrive and progressive abdominal distension since one month of age. There was no associated history of jaundice, fever or bowel complaints. The child looked emaciated and weighed 5.5 kg (< 10th percentile for age). He was anicteric and mild pallor was noted. Abdominal examination revealed fullness on the right side. A soft, large, non-tender lump was palpable in the the right hypochondrium, extending to the right iliac fossa and umbilical region. The lump had a smooth surface and was continuous with liver dullness. Laboratory values were: Hb-10.9 g/dL, total bilirubin-0.7 mg/dL, SGPT-38 IU/L, SGOT-39 IU/L and serum alkaline phospatase-1038 IU/L.

Ultrasound scan of the abdomen and contrast enhanced computed tomography scan revealed a 13 cm ×8.8 cm well-defined round to oval cystic structure occupying almost the entire abdominal cavity with thin peripheral enhancement and continuous with the right and left hepatic ducts. There was no appreciable intra hepatic biliary radical (IHBR) dilatation (**Figures 1a & b**).

The child underwent laparotomy and a GCDC was seen occupying the entire right side of the abdomen, reaching up to the pelvis with the ascending colon, duodenum and pancreas lifted antero-medially. The liver was grossly normal (**Figure 2**). After decompressing the contents (650 mL of greenish bile-tinged fluid) the cyst was completely excised. The common hepatic duct, proximal to the GCDC, was approximately 15 mm in diameter and the lower end of the cyst tapered before joining the pancreatic duct. After flushing the proximal ducts a standard

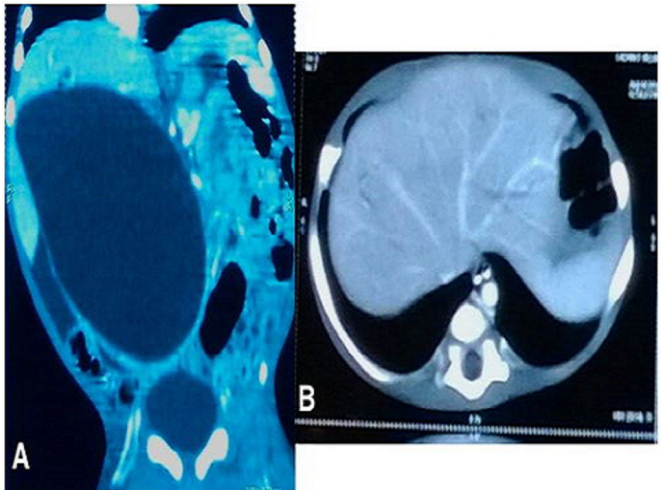

Figure 1: (a) CECT of abdomen in coronal plane showing the giant CDC, and (b): Axial section of the liver showing absence of IHBR dilatation.