A case of Idiopathic Non-Cirrhotic Portal Hypertension (INCPH) in a patient with Klebsiella pneumoniae Pyogenic Liver Abscess (PLA)

Yassine Kilani[1], Dwayvania Miller[1], Ebehiwele Ebhohon[1], Mubarak Yusuf[1], Mohammad Aldiabat[2], Albert Yao[3], Henry Ackerman[3], Anjana Pillai[4]

1: Lincoln Medical and Mental Health Center, Bronx, New York 2: NYU Langone Hospital - Long Island, New York 3: New York Medical College, New York 4 : Morristown Medical Center, New Jersey

BACKGROUND

Pyogenic Liver Abscess (PLA) remains rare in the United States (US). Klebsiella pneumoniae is becoming the leading bacteria in PLA internationally. Causes of bacterial seeding in the liver include biliary tract pathology, portal vein or systemic bacteremia, penetrating wounds or liver surgery. We describe a case of cryptogenic Klebsiella pneumoniae PLA, associated with an incidental finding of idiopathic non-cirrhotic portal hypertension (INCPH).

CASE PRESENTATION

- A 55 year old Hispanic female presented with a week-long history of fever, back pain, jaundice, dark yellow urine and brown feces.
- Our patient was febrile, tachycardic, with scleral jaundice, and a soft and non tender abdomen. Laboratory workup (**table 1**) revealed a pattern of inflammation and cholestatic liver injury. Blood cultures were collected.
- Imaging showed a left hepatic lobe abscess, a heterogeneous hepatic parenchyma, and signs of chronic portal hypertension (PHTN) (**image 1**).
- She was treated with appropriate antibiotics for the liver abscess, and on day 4, CT-guided drainage showed a purulent fluid. Blood and pus culture isolated Klebsiella pneumoniae. A colonoscopy was performed to screen for colonic lesions, and was unremarkable.
- Upper endoscopy found one column of large (>5mm) varices in the lower third of the esophagus which were successfully banded. Screening for causes of liver cirrhosis was negative (including hepatitis B surface antigen and antibody, hepatitis C virus antibodies, iron studies, ferritin, antinuclear, antimitochondrial, anti neutrophil cytoplasmic, anti smooth muscle, and anti-LKM antibodies). Schistosoma antibodies were negative.
- A liver biopsy confirmed the diagnosis of non-cirrhotic portal fibrosis (NCPF) on histology (**image 2**).

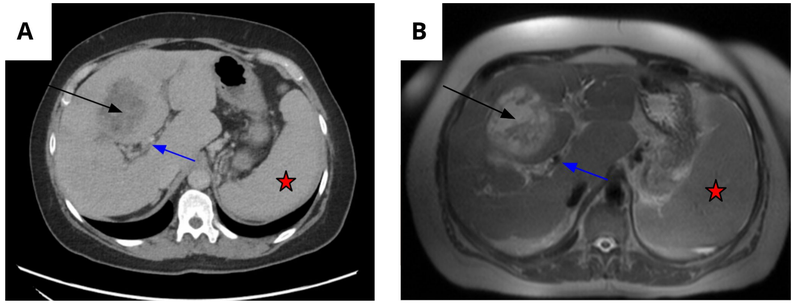

Image 1. Heterogeneous cystic liver mass in the segment IV of the liver measuring 6.2 x 5.8 cm on Computed Tomography (Image A, see black arrow), and Magnetic Resonance Imaging with enhancing internal septa, and enhancing walls (Image B, see black arrow). Additional findings of portal hypertension included a cavernous transformation of the portal vein (see blue arrows), splenomegaly (see red stars) and splenorenal shunt.

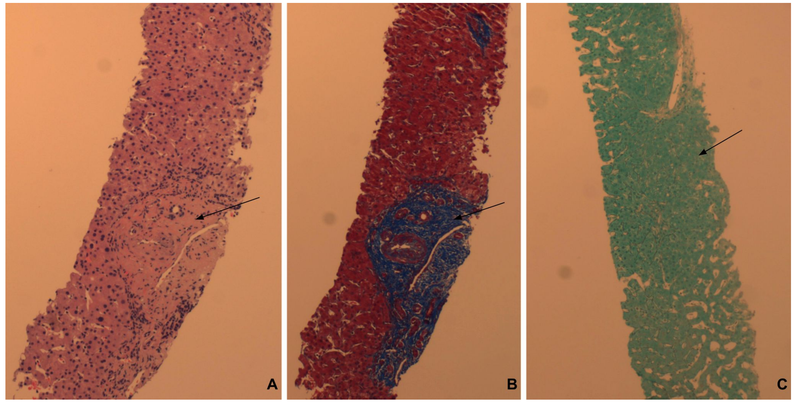

Image 2. Normal liver parenchyma with a few chronic inflammatory cells within portal tracts (Image A, hematoxylin-eosin stain, see arrow), with mild portal fibrosis on trichrome stain (Image B, see arrow), and normal reticulin pattern of hepatic plates on reticulin stain (Image C, see arrow) indicating the absence of liver fibrosis (original magnification X100).

Table 1. Laboratory workup revealing a cholestatic liver injury and inflammation

Lab results	Range & unit	Day 1	Day 9	Day 13
Hemoglobin	12.0 - 16.0 g/dL	10.6	10.5	-
MCV	80.0 - 99.9 fL	89.4	90.0	-
Leukocytes	4.80 - 10.8 .10(3) cells/uL	12.9	8.7	-
Neutrophils	44.0 - 77.0 %	86.8	80.0	-
Platelets	150 - 450 . 10(3)/mcL	180	390	-
AST	≤ 32 U/L	35	-	21
ALT	≤ 33 U/L	49	-	17
ALP	35 - 105 U/L	301	-	230
Total bilirubin	0.20 - 1.20 mg/dL	3.04	-	0.47
Direct bilirubin	0.00 - 0.30 mg/dL	1.90	-	-
Albumin	3.5 - 5.2 g/dL	3.3	-	3.0
PT	10.0- 13.0 seconds	18.7	-	-
INR	mg/dL	1.6	-	-

Discussion

- The incidental discovery of PTHN in patients with underlying liver disease should prompt a full diagnostic workup to rule out liver cirrhosis.
- The diagnosis of INCPH requires each of the following criteria:
  a. clinical signs of PHTN
  b. patent portal veins on imaging (doppler ultrasound or CT)
  c. exclusion of cirrhosis on liver biopsy
  d. exclusion of chronic liver disease that might cause either cirrhosis (Hepatitis B &/or C, non alcoholic steatohepatitis, hemochromatosis, Wilson's disease, primary biliary cirrhosis) or NCPF (Congenital liver fibrosis, sarcoidosis, schistosomiasis).
- INCPH management involves reducing the portal pressure to prevent variceal bleeding and death. Propranolol and endoscopic variceal ligation (EVL) have similar efficacy to prevent rebleeding.