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Case Report

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GIANT, RECURRENT SIMPLE HEPATIC CYST WITH ASYMPTOMATIC PRIMARY BILIARY CIRRHOSIS: AN UNUSUAL ASSOCIATION

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ABSTRACT

The simple hepatic cyst refers to solitary non-parasitic cyst of liver. Simple hepatic cyst is usually asymptomatic but may present as abdominal pain, nausea, vomiting and abdominal distension. These cysts are uncommon with incidence rate of 1% in autopsy study. Hepatic cysts are seen in adults above 40 years of age with female predominance. Primary biliary cirrhosis is a chronic autoimmune cholestatic liver disease with variable clinical course and slow progression. This disease commonly affects females (90%) in the fifth to seventh decades of life. Most of the patients are asymptomatic (>50%) and others present mainly with fatigue (80%) and pruritus (20-70%). Primary biliary cirrhosis is associated with other autoimmune disorders. However to our knowledge, its association with simple hepatic cyst has not been described. We report rare association of a case of simple hepatic cyst with primary biliary cirrhosis in a 55 year old female who presented with abdominal pain and fullness in epigastric region.

KEYWORDS: Hepatic cyst, Primary biliary cirrhosis, Liver.

INTRODUCTION

Hepatic cysts are parasitic or non-parasitic in origin. The non-parasitic hepatic cysts can be classified into four main groups- congenital, inflammatory, neoplastic and traumatic.[1] The term hepatic cyst usually refers to solitary non-parasitic cyst of the liver also known as simple cyst.[1-5] The exact cause of simple hepatic cyst is not known but they are believed to be congenital in origin.[1-4] The cyst is thought to arise as an aberration of bile duct development in utero, which is triggered by chromosome 16.[4]

Primary biliary cirrhosis (PBC) is a chronic autoimmune cholestatic liver disease with variable clinical course and slow progression.[6-9] This disease commonly affects females (90%) in the fifth to seventh decades of life. PBC was first reported by Addison et al in 1851, however Ahrens et al in 1950 coined the term Primary biliary cirrhosis.[6]

We report rare association of a case of simple hepatic cyst with primary biliary cirrhosis in a 55 year old female who presented with abdominal pain and fullness in epigastric region.

To our knowledge this is the first case report of simple hepatic cyst in association with primary biliary cirrhosis.

CASE REPORT

A 55 year old female presented with complaints of pain in abdomen (right hypochondrium and epigastric region) since 1 month. The pain was gradually progressive, dull, aching and non-radiating in nature. No aggravating or relieving factors were associated with the abdominal pain. Patient is a known diabetic and hypertensive currently on treatment. Past history: Patient had history of marsupalisation of hepatic cyst in 2015. Systemic examination: Revealed an immobile, non-tender, swelling in the right hypochondrium and epigastrium with minimal abdominal tenderness on palpation. The laboratory investigations done included Complete Blood Count, RFT, LFT and Serum Amylase (Table.1). The levels of tumour markers were within normal ranges and the indirect hemagglutination test for hydatid disease was negative. CECT Abdomen revealed hepatomegaly with a large well-defined, non enhancing, hypodense cystic lesion in the right lobe of the liver, causing marked stretching and compression of the right branch of the portal vein with hypertrophy of the left lobe (Figure 1a &

1b). Abdominal ultrasonography revealed a large, well defined hypoechoic cystic lesion measuring 19x18x17.8 cm in the right lobe with few septations, and maximum cyst wall thickness of 0.5 cm; causing significant mass effects on liver parenchyma (Figure 1c).

Pathological Findings: Gross examination revealed a single unilocular cystic mass, already cut opened measuring 19x18x17.8 cm. Externally congested, brownish with small bit of liver parenchyma. The wall thickness measured 0.5 cm and inner surface of the cyst was congested, irregularly trabeculated (Figure 2a).

Microscopic Examination: Sections from the cyst revealed cuboidal lining epithelium resting on a fibrocollagenous wall without any cellular atypia or proliferation. These findings are consistent with simple hepatic cyst (Figure 2b). The surrounding liver parenchyma showed features of primary biliary cirrhosis (Figure 2c & 2d) in the form of distortion of liver architecture, creeping fibrosis, lympho-plasmacytic infiltrate surrounding the bile ductules with destruction of bile ductules and within the portal tract, bile ductular proliferation, cholestasis, and sinusoidal dilatation. Focal hepatocytic fatty degeneration was also seen. According to Scheuer's classification, our case corresponds to Stage 3 which is characterized by fibrosis/ scarring.

Table 1: Laboratory Investigations.

Hemoglobin	12 gm%	Serum urea	28 mg%
Total leukocyte count	15,400/ mm^3	Serum creatinine	0.85 mg%
Neutrophil	90%	Total bilirubin	1.13 mg%
Lymphocyte	10%	SGOT	35 IU/L
Platelet count	233x10^3/mm^3	SGPT	26 IU/L
Serum amylase	151 U/L	Alkaline phosphatase	213 U/L

Legends

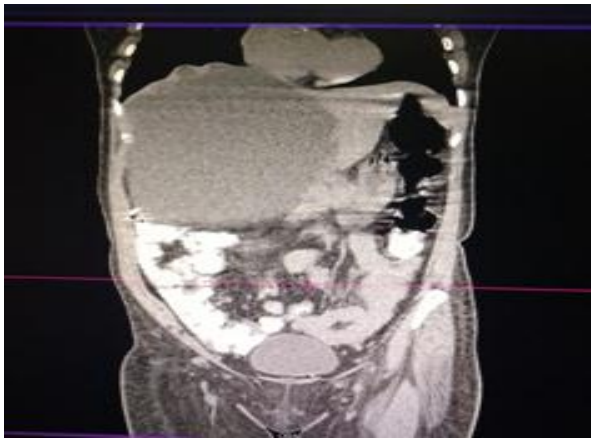

Figure 1a: CECT (Coronal View): A large well-defined, cystic lesion involving right lobe of liver.

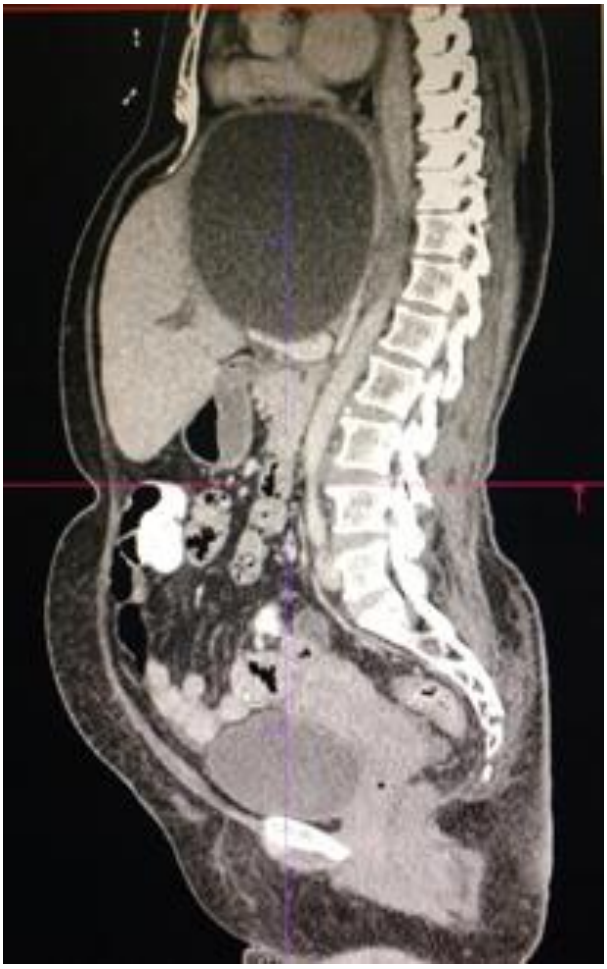

Figure 1b: CECT (Lateral View): A non enhancing, hypodense cystic lesion.

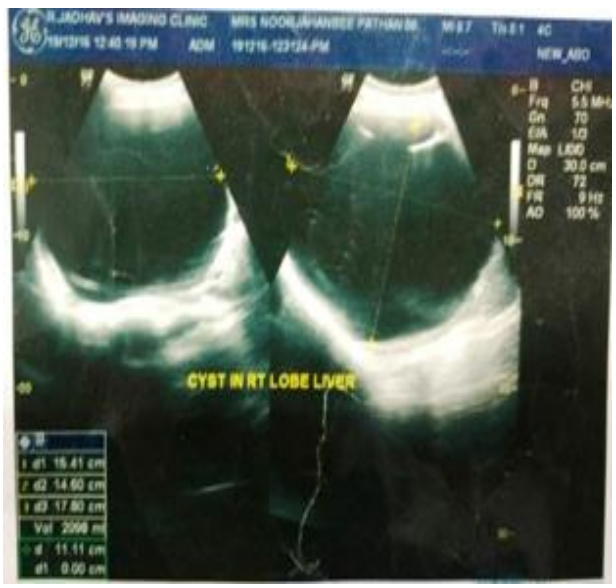

Figure 1c: USG: A hypoechoic cystic lesion in the right lobe of liver with few septations.

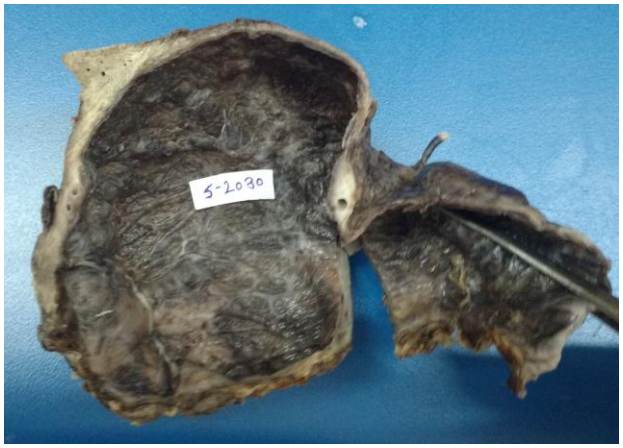

Figure 2a: A 19x18x17.8 cm, unilocular cyst, with trabeculated inner surface.

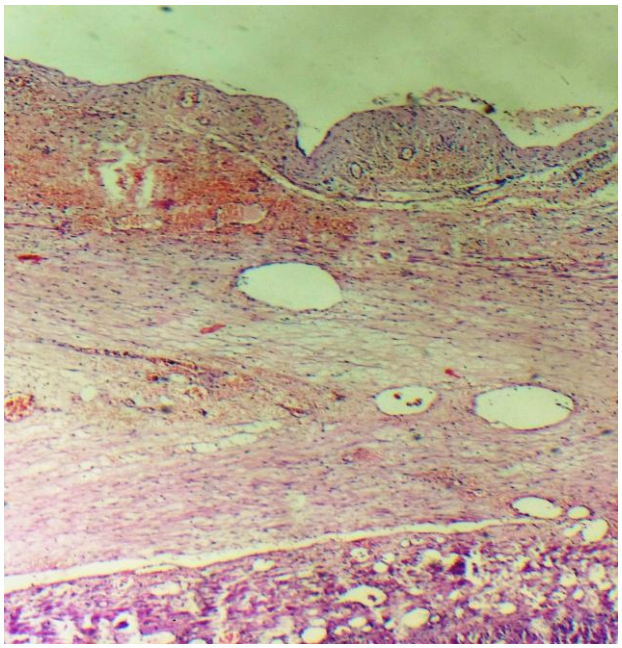

Figure 2b: A cyst lined by cuboidal epithelium with fibrocollagenous wall.

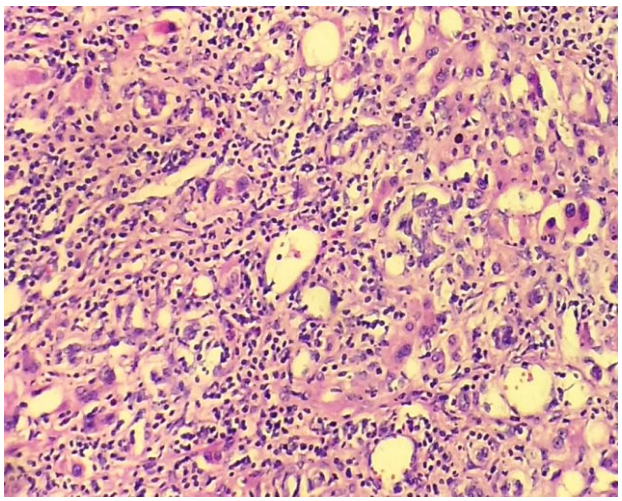

Figure 2c: Microscopy showing features of primary biliary cirrhosis (H&E, X100).

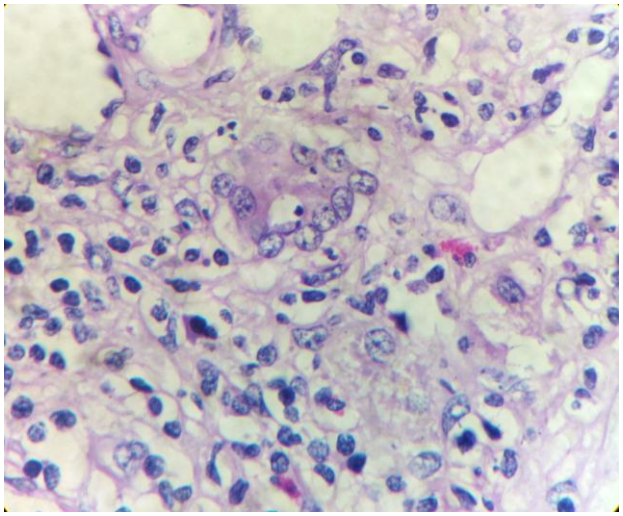

FIGURE 2d: PBC – Chronic non suppurative sclerosing cholangitis (H&E, X400).

DISCUSSION

Simple hepatic cysts are uncommon lesions with a reported prevalence of 1% based on autopsy studies with an estimated prevalence of 2.5% to 18% on imaging-based studies.[2] The cysts are mostly seen in adults (above 40 years) although they can occur in children.[1-4] There is female predominanace with male to female ratio of 1:1.5.[2,4] Simple hepatic cysts commonly involve right lobe of liver, and are small, asymptomatic.[1-5] Large cysts may present with abdominal pain, distension, nausea, vomiting and bloating.[1,3] The giant simple hepatic cysts are uncommon[2,4] and may be complicated as a result of intracystic bleeding, rupture or secondary bacterial infection. Compression of adjacent structures can occur including compression of inferior vena cava.[2,4]

Primary biliary cirrhosis (PBC) is an autoimmune, slowly progressive, cholestatic, liver disease characterized by a triad of chronic cholestasis, circulating anti-mitochondrial antibodies (AMA), and characteristic liver biopsy findings.[6-8] All the three criteria are required for a "definitive" diagnosis of PBC and a "probable" diagnosis requires presence of two of any three criteria.[8] Annual incidence rates range between 0.7 and 49 cases per million population and prevalence rates range between 6.7 and 402 cases per million population.[6,8]

The manifestations and prognosis of this entity differ in patients. PBC is known to progress insidiously through four clinical phases: preclinical, asymptomatic, symptomatic, and liver insufficiency.[9] Patients usually present in fifth to seventh decade and disease is uncommon in patients under 25 years of age.[6-8] Prominent clinical features include fatigue, pruritus, jaundice, xanthomas, osteoporosis, and dyslipidemia.[7,8] Fatigue has been reported in 80%of patients and pruritus in 20 to 70 percent of patients.[8,9] The onset of pruritus usually precedes the onset of jaundice by months to

years.[8,9] Extra hepatic diseases associated with primary biliary cirrhosis include interstitial pneumonitis, celiac disease, sarcoidosis, renal tubular acidosis, hemolytic anemia, and autoimmune thrombocytopenia.[8,9]

PBC is characterised histologically by the asymmetric destruction of the intralobular bile ducts within the portal tracts.[6-9] According to Scheuer's system, Stage 1 is florid duct lesion or chronic non suppurative destructive cholangitis. In Stage 2, there is proliferation of the small bile ductules. Stage 3 is characterised by fibrosis and Stage 4 is cirrhosis.[8,9]

CONCLUSION

The association of simple hepatic cyst with primary biliay cirrhosis is very rare and unusual.

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