Case Report

A rare case of giant biliary mucinous cystadenoma in a 16 year old boy

Monoj Kumar Deka[1*], Anuradha Talukdar[2], Ritesh Tapkire[3], Akalesh K. Verma[4], Ravi Kannan[5]

1Assistant Professor(Pathology), Silchar Medical College and Hospital, Ghungoor, Silchar-788014, India and Visiting Consultant (Pathology), Cachar Cancer Hospital and Research Centre, Meherpur, Silchar-788015, INDIA.
{[2]HOD, Department of Pathology} {[3]HOD, Department of Surgical Oncology} {[4]DBT and Research Associate, Department of Molecular Oncology} {[5]Director} Cachar Cancer Hospital and Research Centre, Meherpur, Silchar-788015, INDIA.
Email: drmonoj_deka@rediffmail.com

Abstract

Introduction: Biliary mucinous cystadenomas of the liver are rare benign cystic tumors constituting less than 5% of intrahepatic biliary cysts. They have recently been redefined as mucinous cystic neoplasms in the 2010 WHO classification. The tumors are insidiously progressive and may attain large proportions. This paper report on a case of giant biliary mucinous cystadenoma in a 16 year old boy. It was 24 cm in size, multiloculated, confined to the right lobe of the liver. Microscopical examination revealed a single lining of cuboidal to tall columnar mucin producing cells with focal stratification. A right hepatectomy was performed. Patient is without recurrence at 3 years follow up.
Keywords: Biliary mucinous cystadenomas, giant, case report, Histopathology.

[*]**Address for Correspondence:**
Dr. Monoj Kr Deka, Visiting Consultant, (Pathology), Cachar Cancer Hospital and Research Centre, Meherpur, Silchar-788015, INDIA.
Email: drmonoj_deka@rediffmail.com
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INTRODUCTION

Biliary mucinous cystadenomas of the liver are rare benign cystic tumors constituting less than 5% of intrahepatic biliary cysts[6,21]. In the 2010 WHO classification they have been redefined as mucinous cystic neoplasms[23]. The exact etiology of the tumor is unknown. An anomalous development of the primitive hepatobiliary analage may be responsible for its origin[2,3]. The tumor may attain large proportion and prone to malignant degeneration[6]. This paper reports on a case of giant biliary mucinous cystadenoma in a 16 year old boy.

MATERIAL AND METHODS

The present study on a case report of Giant biliary mucinous cysadenoma was undertaken in Cachar Cancer Hospital and Research Centre, Meherpur, Silchar, India.

CASE REPORT

A 16 year old boy was admitted with complaints of a dull ache and an enlarging mass in the right upper abdomen of 10 years duration. Patient was thin built with very large cystic mass almost occupying entire abdomen. Seconday sexual features were absent. Patient had history of laparotomy 4 years back. Previous record showed that the hepatic cyst was marsupialized and omentum was placed in the cyst cavity. Liver function tests and full blood count were within normal limits. Viral markers for hepatitis B and C were negative. Ultrasound followed by CT scan revealed a large multilocular cystic lesion with internal septa occupying the abdominal cavity predominantly on the right side and extending inferiorly up to the pelvis displacing the adjacent structures to the left. In view of previous history, the diagnosis was recurrent hepatic cyst with possible malignancy in view of large size. Ultrasonographic guided fine needle aspiration cytology from the mass demonstrates the cystic nature of the lesion. The patient underwent laparotomy. The cyst was arising from right lobe of liver, and adherent to hilar structures, IVC displacing bowel loops to left of

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abdominal cavity. Right hepatectomy was done. Tear in left hepatic duct was repaired with placement of internal stent. Re-exploration was done on 2nd postoperative day due to secondary haemorrhage but no major bleeding was found except for some minor ooze from peritoneal and retroperitoneal raw areas. Rest of the postoperative period was uneventful. Histopathology Report

Gross examination showed a large cystic mass occupying almost the whole of right lobe of liver measuring 24 cm in diameter with small amount of normal liver tissue adherent to the cyst wall. Cut surface showed multiloculation containing fluid and mucin. Multiple sections from the cyst wall showed single lining of cuboidal to tall columnar mucin producing cells with focal stratification. Underlying wall showed congestion and chronic inflammatory infiltrate predominantly plasma cells and lymphocytes. There was no evidence of cellular pleomorphism or invasion into underlying stroma. Adjacent liver tissue was unremarkable. Features were consistent with giant biliary mucinous cystadenoma. Biliary stent was removed endoscopically on follow up. At 3 years follow up, patient is disease free, with significant weight gain and there is development of secondary sexual features. Ultrasonography revealed enlargement of left lobe of liver.

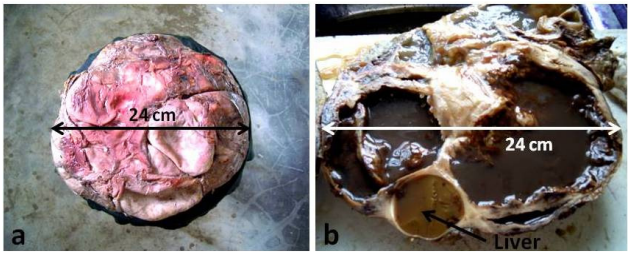

Figure 1

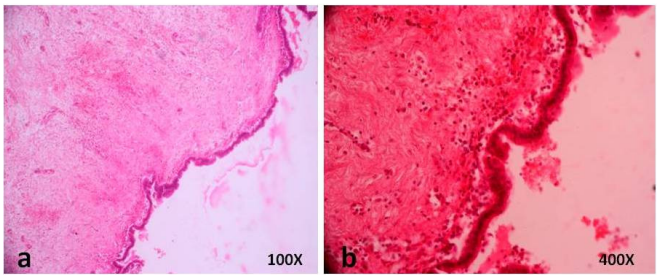

Figure 2

DISCUSSION

Biliary mucinous cystadenomas are rare benign cystic tumors, derived from the biliary epithelium. Predominant location is the liver especially right hepatic lobe, less frequently in the extrahepatic bile duct and rarely in the gall bladder[4,15, 19,20]. They occur almost exclusively in middle aged women 40 to 50 years[7,16,20]. Clinical features are non specific. Most of the patients present with a sense of local fullness and a visible distension. The duration of symptoms may vary from a few weeks to as long as 12 years [2,7,10]. Other clinical manifestation include jaundice, infection and abdominal pain[13]. This case highlights the exceptional occurrence in a adolescent boy. Serum CA19-9 may be elevated but high level is inconsistent with the lesion[5]. Radiological studies including ultrasonography and CT scan may be helpful in delineating the tumor; but these do not provide specific diagnosis[2]. Biliary mucinous cystadenomas are appreciated as premalignant lesions[1,17,18]. Radiological studies and tumor markers are not significant for differentiating cystadenoma and cystadenocarcinoma[1,5,17]. Therefore histopathology is essential to confirm the diagnosis and more important to exclude malignancy. Macroscopically biliary cystadenomas are multilocular cysts containing clear to mucinous fluid. The size varies from 1.5 to 35 cm [21,24,25]. In rare cases the content of the fluid may be bile stained, purulent, proteinacious or gelatinous[21,22]. A single layer of biliary-type, cuboidal to tall columnar, non ciliated, and mucin secreting epithelial cells with papillary projection lined the cyst wall[4,13] Exclusively in women the tumor may contain an ovarian like stroma[8,9]. In our case the size reached huge proportion upto 24 cm in diameter during a 10 year period. Because of the malignant potential and risk of recurrence, complete excision with negative margins is the treatment of choice[5,7,11,14]. Aspiration and sclerosing therapy, marsupialization, and internal drainage should be avoided because of high recurrence rate of the tumor[12]. In this case complete excision is associated with excellent outcome.

CONCLUSION

We are reporting this case because of its rare incidence. Histopathology is crucial in confirming specific diagnosis and ruling out malignancy. Complete excision is the optimal treatment.

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