

## Biliary Cystadenoma Causing Esophageal Varices

Sung Ju Kang, Tae Hee Lee, Min Gyu Seok, Hyo Jin Yun, Ye Seul Jang, Jun Hyun Byun

*Department of Internal Medicine, College of Medicine, Konyang University, Daejeon, Korea*

Biliary cystadenomas are benign but potentially malignant cystic neoplasm. The preferred treatment is radical resection because it is difficult to differentiate a benign from a malignant biliary cystadenoma. A 40 year-old woman presented with moderate abdominal discomfort. Esophageal varix was found up to mid-esophagus on endoscopy. She has no prior history of liver disease or chronic alcohol ingestion. About 15cm sized biliary cystadenoma was diagnosed by ultrasonography, computed tomography and magnetic resonance imaging. Serum level of bilirubin, alanine aminotransferase, alkaline phosphatase, gamma-glutamyl transpeptidase and tumor marker were elevated. The patient underwent US-guided aspiration. Tumor markers from the aspirated fluid are increased. Left hepatectomy was performed to completely remove the cyst. Histology of the resected specimen confirmed a biliary cystadenoma of the liver with ovary-like stroma. Without prior history of liver disease or chronic alcoholic ingestion, incidental finding of esophageal varix could show an important clue for diagnosis of biliary cystadenoma.

**Key Words:** Biliary, Cystadenoma, Esophageal varix

Biliary cystadenomas are benign but potentially malignant cystic neoplasm. They are rare multilocular cystic tumors of the liver that usually located in the right hepatic lobe. The incidence is less than 5% of intrahepatic cyst. Biliary cystadenomas present predominantly in middle-aged women.<sup>1</sup> Radiological imaging, often abdomen computed tomography (CT), is fundamental in diagnosis. On CT image, biliary cystadenoma appears as a low attenuated mass, which may be uni- or multilocular, or may have septations. The preferred treatment is radical resection because it is difficult to differentiate a benign from a malignant biliary cystadenoma.<sup>2</sup> Some

patients present abdominal pain, nausea, vomiting, and obstructive jaundice. But, often they are asymptomatic. In some case biliary cystadenoma incidentally found during physical health examination. However, it is difficult to detect cystic tumor unless symptoms are presented. The lesions were incidentally found on abdominal images such as abdomen ultrasonography (US) or abdomen CT. But it is rare to associate esophageal varix with biliarycystadenoma.

### CASE

In September 2013, a 40 year-old woman pre-

**Corresponding Author:** Tae Hee Lee, Division of Gastroenterology and Hepatology, Department of Internal Medicine, Konyang University College of Medicine, 158, Gwanjeodong-ro, Seo-gu, Daejeon 35365, Korea Tel: +82-42-600-9370 Fax: +82-42-600-9090 E-mail: green740@naver.com

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sented with several week history of moderate abdominal discomfort. But the patient's general health was good and she had no history of any liver disease. She visited a local hospital for routine check up. Clinical examination was normal. When she underwent an endoscopy, esophageal varix was found upto mid-esophagus. Abdominal US confirmed 15 × 10 cm cystic mass in the liver (Fig. 1). Cystic mass appears as a hypoechoic lesion with multiple thin-walled septae. Abdominal CT confirmed presence of 16.2 cm sized cystic lesion with internal septation (Fig. 2).

Serum levels of bilirubin (1.58 mg/dL), alanine aminotransferase (224 IU/L) alkaline phosphatase (369 IU/L) and gamma-glutamyl transpeptidase (1182 IU/L) were elevated. Tumor marker Carbohydrate antigen 19-9 (44.7 U/ml) were elevated. She admitted to our hospital for further evaluation. Magnetic reso-

nance imaging (MRI) showed that the 16 cm sized cystic lesion with thin-walled and irregular enhancing septae. Both intrahepatic ducts were dilated due to extrinsic compression by the cystic lesion. To differentiate the cystic mass benign from malignancy, she underwent US guided aspiration. Cytology of fluid showed histiocytes and necrotic debris. Tumor marker are increased which were analyzed from aspiration fluid (Carcinoembryonic antigen 103.24 ng/mL, Carbohydrate antigen 19-9 > 1900 U/mL). The patient was scheduled for surgery based on the ultrasound, CT, MRI and cytology findings. Left hepatectomy was performed to completely remove the cyst. During operation, clear mucinous fluid were shown and cyst wall was resected for frozen section (Fig. 3). Histology of the resected specimen confirmed a biliary cystadenoma of the liver with ovary like stroma (Fig. 4).



Fig. 1. Abdomen ultrasonography shows a large cystic mass with septae in liver left lobe.

Two months after surgery, she had been well. Follow up CT showed complete resection of cystic lesion and no evidence of recurrence. Follow up endoscopy revealed complete disappearance of esophageal varix (Fig. 5).

## DISCUSSION

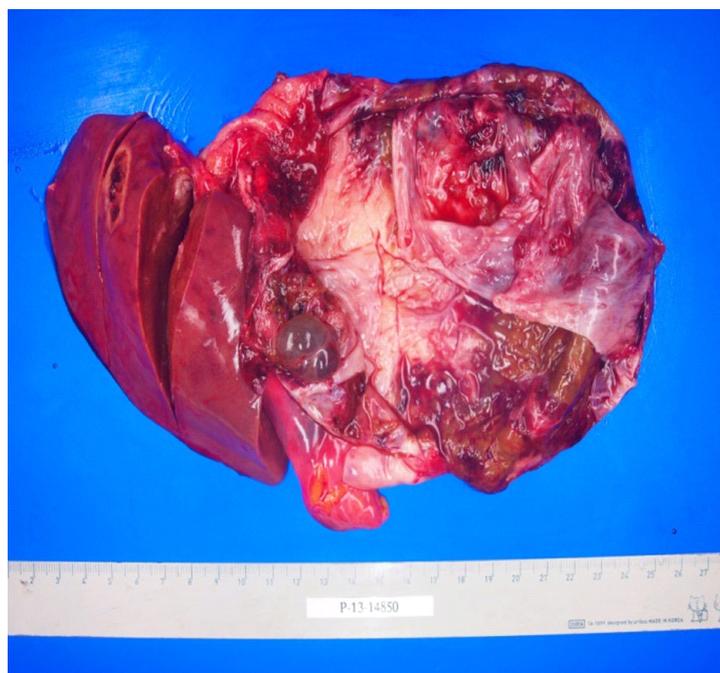
Biliary cystadenomas are rare cystic neoplasm that usually arise in the liver or extrahepatic bile ducts and may show malignant degeneration into cystadenocarcinomas. The peak frequency of cystadenomas is in 30 - 50 years old women.<sup>3</sup> Biliary cystadenomas are often asymptomatic. The clin-

ical presentation may occur when the tumor reach a significant size and cause compression of adjacent structures. Imaging studies by US, CT and MRI play an important role in confirmation of the lesion. US help to differentiate cystadenoma from simple hepatic cyst. On US, biliary cystadenoma usually showed a hypoechoic lesion with thick-end, irregular walls. CT reveals a hypodense, thick walled cystic mass with internal septa. MRI can give information of cystic fluid. Cystic lesion appears hyperintense on T2-weighted images and hypointense on T1-weighted images.

On histopathological examination, biliary cystadenoma can be divided into 2 groups. Two groups can be distinguished according to the



Fig. 2. Computed tomography shows a large cystic lesion with multiple-enhancing septae (arrow).



**Fig. 3. Postoperative picture shows large thick walled cystic lesion in left lobe of liver.**

presence or absence of mesenchymal stroma, ovarian stroma. The lining of mucinous biliary cystadenoma is microscopically composed of mucus-secreting cuboidal or columnar epithelium. This tissue expresses receptors for female sex hormone (estrogen and progesterone). Biliary cystadenomas tend to occur predominantly in women because of hormonal responsiveness. Some studies showed pathological correlation between ovarian stroma originating in liver, pancreas, or retroperitoneum and mucinous neoplasm of the ovary. In embryonic period, the epithelial cells of the gonads detach from their tissue boundaries and migrate into the surface of nearby organ such as liver. Biliary cystadenoma without ovary-like stroma does not stain for estrogen and progesterone receptors.<sup>4</sup>

The treatment of choice for biliary cystadenoma

is radical resection. No medical treatment has been found to be effective. Complete resection should be performed whenever possible since it is believed to be premalignant. Aspiration or partial excision is associated with higher recurrence rate. Gambelin et al, investigated the efficacy of laparoscopic resection of hepatic cysts.<sup>5</sup> According to histologic examination, 90% of the lesions in the study were simple cysts, and 10% of them were cystadenomas. All patients who were operated on for pain experienced symptom relief. Incidental findings of esophageal varix need to be considered another provoking factor of portal hypertension. In our case, an asymptomatic patient who incidentally found esophageal varix in routine endoscopic examination with increased liver enzymes. After operation, esophageal varix disappeared at follow-up endoscopy with normalized liver enzymes. It

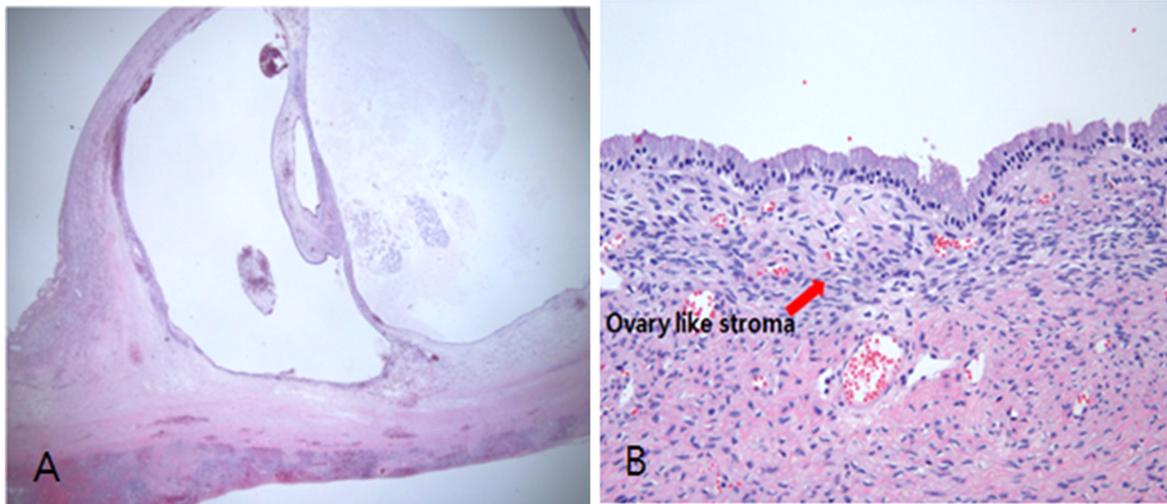


Fig. 4. Cyst histology typical of a mucinous cystadenoma(A) Biliary cyst shows ovary like stroma (H & E stain,x400). (B) Arrow demonstrating ovary like stroma.

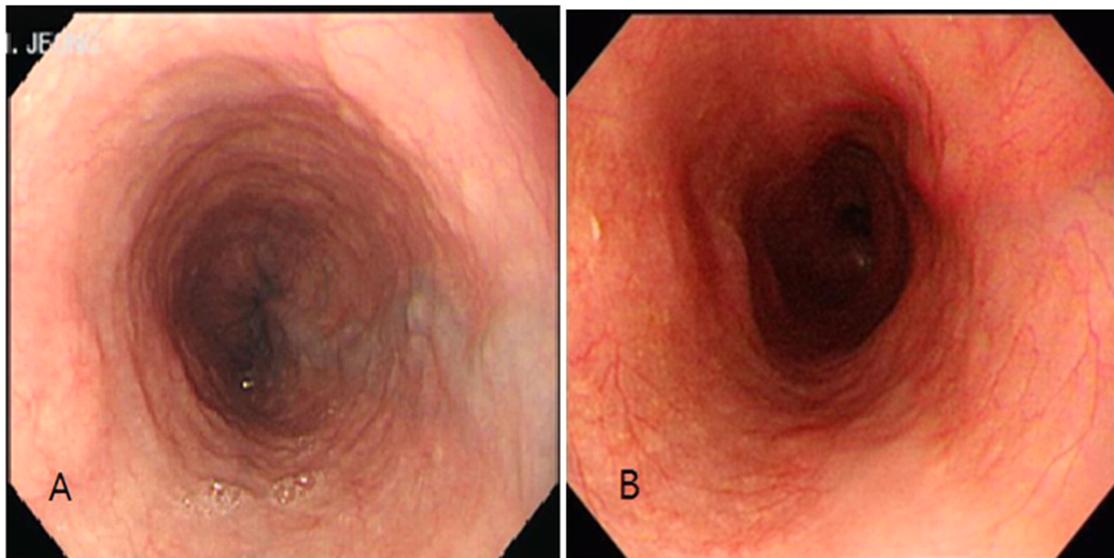


Fig. 5. Endoscopy image before operation demonstrating esophageal varix upto midesophagus.(A) Follow up endoscopy after lobectomy shows disappearance of esophageal varix.

is important to diagnosis biliary cystadenoma before transformed to malignant. Imaging studies by US, CT and MRI play a important role of detecting lesion. However, like our study, incidental finding of esophageal varix and elevated CA 19 - 9 and CEA in cystic fluid can show important clue of diagnosing biliary cystadenoma.<sup>6</sup> Without prior history of liver disease or chronic alcoholic ingestion,

incidental esophageal varix imply considerable size hepatic or biliary mass or vascular lesion which induced portal pressure increasing.<sup>7</sup>

Biliary cystadenoma are potentially malignant cystic neoplasm. Complete resection is the best treatment. It is difficult to detect cystic tumor unless symptoms are presented. Imaging studies by US, CT and MRI play an important role of detect-

ing lesion. However, without prior hx. of liver disease or chronic alcoholic ingestion, incidental finding of esophageal varix can show important clue of diagnosing biliary cystadenoma.

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