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Biliary Cystadenoma Rupture: A Case Report of Emergency Laparoscopic Treatment

Bilier Kistadenom Rüptürü: Acil Laparoskopik Tedavi Uygulanan Bir Olgu Sunumu

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ABSTRACT
Biliary cystadenoma is a rare cystic lesion of the liver and acute abdomen caused by perforation of the cyst is very unusual. We report a case with rupture of biliary cystadenoma (with a diameter of 25 cm) localized in the right lobe of the liver, which we diagnosed during laparoscopic marsupialization and planned to resect with a second operation.
Key Words: Biliary cystadenoma, ultrasonography, laparoscopy, rupture

ÖZET
Bilier kistadenom karaciğerin seyrek görülen kistik lezyonudur ve perforasyon gibi komplikasyonu nedeni ile akut abdomene sebep olması çok nadirdir. Biz de karaciğer sağ loba lokalize 25cm çapında, rüptüre olmuş, laparoskopik marsupiyalizasyon ile tanısı konulan ve ikinci bir operasyonla rezeksiyonu planlanan bilier kistadenom vakasını sunuyoruz.
Anahtar kelimeler: Bilier kistadenom, ultrasonografi, laparoskopi, rüptür

INTRODUCTION

Biliary cystadenoma is a rare cystic lesion of the liver, derived from bile epithelium, and is very hard to diagnose in the preoperative period (1). It accounts for more than 5% of hepatic cysts and 1% of hepatic cystic lesions (2). Biliary cystadenoma is mostly observed in middle-aged women, with a diameter range of 1.5-35cm (3). Lesions, which are usually asymptomatic, are detected on radiological evaluations for other clinical indications or they are discovered incidentally during surgical exploration (3). Initial findings of cystic lesions, such as massive hepatomegaly, bleeding, infection, jaundice and rarely vena cava inferior obstructions, are observed generally in the 5th decade (2).

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Although biliary cystadenomas are benign lesions, they have a tendency for recurrence, and there is risk of malignant progression (2,3). Clinical characteristics and effective surgical treatment of these lesions, which

may be mistaken for echinococcus cysts and simple cysts, have not been clearly determined. Worldwide prevalence of biliary cystadenomas, which have been reported in less than 200 cases in the literature, is unknown (3,4). Acute abdomen caused by complications due to perforation of cysts is very rare. We present a case with rupture of biliary cystadenoma (with a diameter of 25 cm) localized in the right lobe of the liver, which we diagnosed during laparoscopic marsupialization and planned to resect with a second operation.

CASE REPORT

Ultrasound evaluation in a 30-year-old woman in the 2nd month of pregnancy incidentally detected a cystic structure with a diameter of 22cm covering the right lobe of the liver. There were multiple septations and formations such as daughter cysts inside the cyst (figure 1). Tomography could not be applied because of pregnancy. Follow-up was found appropriate after echinococcus hemagglutination test was determined to be negative. In the 3rd month of pregnancy, the patient developed pain in the right upper quadrant. Palliative decompression of the cystic lesion and diagnostic aspiration were performed using ultrasound guidance. Histological evaluation of the light yellow-clear fluid revealed no signs of echinococcus infection or malignancy of epithelial origin. As there was no membrane detachment during the aspiration and the aspirate did not resemble "eau de roche", a pre-diagnosis of hydatid cyst was excluded. As possibilities of hydatid cyst and malignancy became less likely, treatment of the cyst was planned to be performed electively six months following the end of pregnancy.

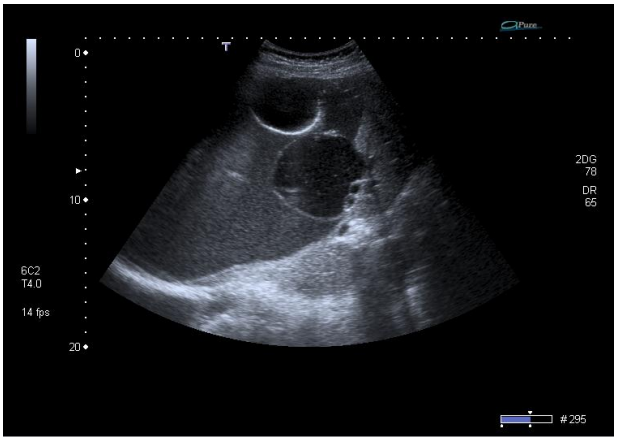

Figure 1: Multiple septations and formations such as daughter cysts inside the cyst.

In the 3rd month postpartum, the patient presented to emergency services with diffuse abdominal pain with sudden onset and increasing intensity. The patient had no complaint other than abdominal pain and her medical history was unremarkable except for the cesarean operation three months before. On physical examination, there were tenderness, defense and rebound in all quadrants but there was no palpable mass. There were no pathological findings in laboratory evaluations (CRP: 2.18mg/dL; white blood cell: 7700/mm^3). Ultrasound evaluation showed a cystic structure (24x18x12cm) with irregular borders covering the right lobe of the liver, and there was anechoic free fluid with depths of 5cm in perihepatic area and 3cm in pelvic area (figure 2). Exploration of the case during emergency laparoscopy, with a pre-diagnosis of acute abdomen due to liver cyst rupture, revealed a ruptured simple cyst (25x20cm) in the right lobe of the liver and clear free fluid in the abdomen. Although diagnosis of hydatid cyst had been excluded, precautions were taken and marsupialization was performed to the cyst wall at the border of the healthy liver tissue. Free fluid in the abdomen was aspirated and abdominal washing was performed with warm serum physiologic. At the end of the two-hour operation, the abdomen was desufflated with two drains. Drains were removed on postoperative day 2 and the patient was discharged. Histopathological examination of the specimen showed cystic lesion with a stroma of ovary-like fibroblasts with spindle-like nuclei, and one-layer mucinous epithelium. Immunohistochemically, stromal cells were stained with estrogen, progesterone receptors and alpha inhibin, whereas cyst epithelium was stained with MUC-2, and mucinous type bile duct cystadenoma was reported.

Cyst resection with a second operation was planned after completion of the lactation period.

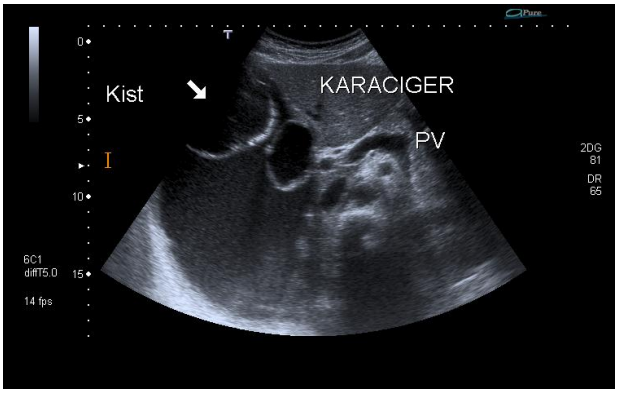

Figure 2: Cystic structure with irregular borders, covering the right lobe of the liver.

DISCUSSION

Cystic liver lesions are classified into developmental, neoplastic, inflammatory and miscellaneous types (5). Benign cystadenoma and cystadenocarcinoma are cystic neoplasms originating from intrahepatic bile epithelium (90%), and they constitute less than 5% of intrahepatic cystic lesions (6). They rarely originate from extrahepatic bile ducts or gallbladder (3). After the first case was defined by Keen in 1892, Edmonson et al. was the first to define multiloculated cysts with pseudo-ovary stroma and mucus-secreting ductal epithelium, in 1958 (4). Incidence of biliary cystadenoma is 1/20,000-100,000, while incidence of cystadenocarcinoma is 1/10,000,000 (3). The etiology of biliary cystadenoma is unknown. The tumor is thought to result from the development of ectopic rests of primitive foregut sequestered within the liver or to be due to the obstruction of the congenitally aberrant bile duct (7). Of all cystadenomas, 50-55% are observed in the right lobe, 30-40% in the left lobe and 15-20% in both lobes (3).

Most of the lesions are seen in middle-aged women (80%), supporting the role of the hormonal effect (2-4), and occur between the 4^{th} and 6^{th} decades (2). Female gender of the present case and growth of the cyst to a symptomatic state during the pregnancy support the hormonal effect.

Biliary cystadenoma is usually detected incidentally by screening techniques such as tomography and ultrasound (2,3). In symptomatic cases, there are nonspecific complaints such as abdominal pain, abdominal distension, and palpable epigastric mass or epigastric distress. If complications such as secondary cyst infection, tumor perforation, jaundice, cholangitis or intracapsular bleeding occur, early diagnosis may be possible (2). As in this case, patients may present to emergency service because of acute abdomen symptoms due to cyst rupture. The number of cases operated on an emergency basis due to biliary cystadenoma perforation is very low. In our case, laparoscopic marsupialization was performed for perforated cystadenoma in the right lobe of liver and the nature of the lesion was determined by histopathological evaluation. The following should be considered in the differential diagnosis of hepatic cystic lesions: simple cysts, complicated cysts (hemorrhagic or infected cysts), bilomas, hematomas, abscess, echinococcus cysts, cystadenoma or cystadenocarcinoma, cystic hamartomas, cystic-necrotic neoplasias, peliosis, polycystic liver disease and Caroli disease (2,4). With the help of anamnesis and other clinical-screening and laboratory findings, most of these may be excluded, but radiological findings may not always be sufficient for differential diagnosis of simple and hydatid cysts, in addition to biliary cystadenoma and adenocarcinoma (8).

In our case, biliary cystadenoma was considered based on histological findings after the aspiration by ultrasound and final diagnosis was only possible after the histopathological evaluation of the material obtained during laparoscopic approach.

Radiological findings are important in the differential diagnosis of hepatic cystic lesions. Especially cyst content, septations and details of the cyst wall can be evaluated well with ultrasound and the cyst character may be described (4). There are no thick septa, intense content or thick-calcified wall in simple cysts. For hydatid cysts, the wall is thick and wall calcification frequent, and membrane detachment, daughter cysts, hydatic sand and septa may be helpful in the diagnosis for some types. However, septations and structures such as daughter cysts may also be observed in biliary cystadenoma and cystadenocarcinoma. In such cases, serology and cytological analysis of the fluid obtained by aspiration are necessary. Mural nodules in the cyst wall and vascular signals from these nodules by Doppler ultrasound are pathognomonic for biliary cystadenocarcinoma. If no mural nodules are present, histological evaluation is necessary for certain diagnosis.

Radical excision of the cystic lesion should be the first preferred treatment (9). Although biliary cystadenomas are benign lesions, they have a high frequency of recurrence and potential for malignant transformation (3,10). The malignant transformation rate is high, reported as more than 30% (2,4); thus, aspiration and sclerosing treatment, marsupialization and internal drainage should not be performed (2,3). Recurrence is observed at a rate of two-thirds in patients who undergo only local or pericystic excision, whereas 10% of hepatectomy cases may even require additional surgical approach due to recurrence (10). Recurrence rate is up to 90% in cases of incomplete resection (3).

Hence, for patients with cyst of the liver, it is important to perform appropriate radiological screenings, to establish certain diagnosis, and to give the appropriate treatment, since prognosis of cystadenoma is very good in patients whose lesions are removed totally. There are few publications in the literature about successful laparoscopic treatment of biliary cystadenoma (3). Two-stage surgical treatment may be performed in cases who become symptomatic with acute clinical condition and in those misdiagnosed in the preoperative period (2). In the case presented here, a two-stage operation was performed. First, laparoscopic marsupialization was applied to the patient presenting to the emergency service with acute abdomen, the diagnosis of biliary cystadenoma was ascertained based on histopathological evaluation of the cyst wall, and pressure symptoms due to the huge cyst and rupture complications were eliminated. After pathological diagnosis of the biliary cystadenoma, we have planned to resect the remnant cyst by 2 cm around the capsule.

Biliary cystadenomas may cause acute abdomen due to complications such as perforation. When huge cysts in the liver are detected radiologically, especially in women in their 3rd or 4th decade, biliary cystadenoma must be considered in the differential diagnosis. In these lesions, which may be mistaken in the preoperative period for simple cysts, hepatic abscess or echinococcus cysts, laparoscopy may be performed for differential diagnosis. In cases in whom biliary cystadenoma diagnosis is ascertained, total excision of the cyst, enucleation or hepatic resection should be performed due to the high malignant transformation and recurrence risk.

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