

Biliary Cystadenoma Causing Obstructive Jaundice: Case Report and Literature Review

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Key Words

Hepatectomy · Cyst · Liver

Abstract

Biliary cystadenomas are rare, potentially malignant neoplasms of biliary origin. Presentation is usually with vague and non-specific symptoms. Here, we describe an unusual case of biliary cystadenoma in a woman presenting with acute onset obstructive jaundice and review the relevant literature of 26 such cases reported over the last two decades.

Case Report

A 40-year-old, previously fit, Asian woman presented with obstructive jaundice. There was no significant history except for recent travel to Pakistan. Physical examination was normal apart from features of obstructive jaundice. Liver function tests revealed elevated serum alkaline phosphatase (419 IU/l), alanine transaminase (253 IU/l) and bilirubin (50 µmol/l). Hepatitis and hydatid serology were negative. Abdominal ultrasound (US) showed a 7 cm diameter cystic lesion in the region of the porta hepatis with an irregular echo pattern and dilatation of the left hepatic duct. Magnetic resonance cholangiopancreatography (MRCP) and CT confirmed a septate cystic lesion situated close to the porta hepatis ([fig. 1](#), [fig. 2](#)); wall calcification or enhancement was not observed. Subsequent endoscopic retrograde cholangiopancreatography (ERCP) delineated a stricturing, filling defect at the junction of left and main hepatic ducts, corresponding to the area seen on cross-sectional imaging. Biliary brushings were negative and biliary stenting unsuccessful, prompting surgical exploration after discussion at the multi-disciplinary meeting. A left hepatectomy was performed and a polypoid mass lesion contiguous with the main hepatic cystic lesion was seen entering the main hepatic duct from the left hepatic duct. The left hepatic duct was transected at the confluence and the rest of the biliary tree was confirmed as normal by choledochoscopy prior to its closure. Gross pathological examination of the specimen revealed a thick walled cyst obstructing the left hepatic duct ([fig. 3](#)), and histology confirmed a biliary cystadenoma. Follow-up for 1 year after was uneventful.

Literature search using free text search 'biliary cystadenoma' and 'obstructive jaundice' along with related article search function and hand-searches of retrieved articles found 26 similar cases published

over 24 articles from 1984 to 2008. In this article we review the clinical course of the patients described in the last 3 years (2006–2008).

Discussion

Biliary cystadenomas are rare, potentially malignant neoplasms of biliary origin occurring predominantly in middle-aged women [1]; the first description of their resection was by William Williams Keen in the 1890s [2]. Biliary cystadenomas are composed of communicating, variable sized locules usually containing clear fluid. The locules are lined by simple, predominantly columnar epithelium resembling biliary epithelium with cytoplasmic mucin. Typically, there is cellular mesenchymal tissue resembling ovarian stroma. The epithelium may show varying degrees of dysplasia. High grade dysplasia or invasive carcinoma suggest transformation to cystadenocarcinoma [1]. The cyst size at presentation is usually between 5 and 15 cm, its expansion causing vague, non-specific upper abdominal symptoms, such as mass, sometimes mimicking biliary calculous pain or gastroduodenal (acid-peptic) symptoms such as nausea [3–12]. Jaundice is an infrequently observed clinical feature caused by obstruction of biliary flow, usually transient and due to direct external compression of the biliary tree. Usually no direct communication exists between the cyst and the biliary tree although occasionally cases investigated with ERCP have demonstrated cyst filling [5]. Rarely, progressive or intermittent jaundice may be observed, as noted in our case, due to polypoid extension into the bile duct causing ball-valve obstruction, without evidence of invasion in a benign lesion. Previous reports of biliary cystadenoma presenting with obstructive jaundice are summarized in [table 1](#).

In the past, ultrasound has been reported to be at least as sensitive as CT at demonstrating septa, when taken in the context of the history to suggest cystadenoma or cystadenocarcinoma [10]. CT supplies further information that helps differentiate these two diagnoses by revealing any protruding nodules, significant wall enhancement or calcifications, typical of malignant cystadenocarcinoma [10]. Recent reports have inferred advantages of biopsy or aspiration from cysts in the hope of identifying malignant change preoperatively [7]. Routine use cannot be advised due to risks of carcinomatosis, cyst rupture and the high probability of missing areas demonstrating atypia due to septation [5]. Interestingly, cystadenocarcinoma is encountered more commonly in men (than cystadenoma) resulting in only a slight propensity for women [11]. Additionally presentation is apparently later in life at a mean age of 10 years after that of cystadenoma [11]. Weight loss and symptoms of metastases may contribute to the clinical picture. Fortunately the ratio of cystadenocarcinomas to cystadenomas is low.

Hepatic cysts are noted in 1–5% of the general population, with the most common pathologies being simple cysts, cystic necrosis of tumours, hydatid cysts, choledochal cysts, biliary cystadenoma amongst others. Simple hepatic cyst is extremely common, incidentally noted in 18% of patients investigated for unrelated pathologies [12]. Patients are managed by marsupialization, usually laparoscopic, only when symptomatic to achieve a more lasting cure as opposed to simple percutaneous aspiration. A hydatid cyst caused by the tapeworm *Echinococcus granulosus* or *Echinococcus multilocularis* is suspected when synchronous cysts are found (in 75% of infections) in addition to findings of eosinophilia and frequently with a history of inhabitation in an endemic area. CT findings of these and some less common differentials are provided in [table 2](#).

Histological examination is required before a definitive diagnosis can be made. Factors such as bleeding within a benign locule, especially after attempted manipulation, can

cause confusion with cystadenocarcinoma at imaging. Optimum management of cystadenomas involves either hepatectomy or, at the very least, enucleation. For a fully resected cystadenoma prognosis is excellent, recurrence being uncommon [9]. Where resection is subtotal for anatomical (biliary or vascular) reasons, it is recommended patients are followed up at least yearly for the first 2 years as recurrence is said to be most likely during this period [13]. Rare pathological variants may include serous type, which may regress spontaneously. Dysplasia or neoplasia may involve only part of the cyst wall [14].

Conclusions

(1) Cystic lesions of the liver may pose a diagnostic dilemma. Simple cysts are by far the most common lesions encountered. (2) Needle aspiration is not recommend for preoperative diagnosis. (3) Imaging in conjunction with a good history is usually sufficient to distinguish cystic hepatic lesions. (4) Jaundice can occur even in benign lesions, without signs of invasion, due to direct compression or ball-valve obstruction of the biliary tree by the cyst.

Table 1. Previous reports of biliary cystadenoma presenting with obstructive jaundice

Reference	Demographics (No; age; sex)	Radiology	Management	Follow-up
This report	1; 40; F	US, CT, ERCP, MRCP	left hepatectomy	12 months
Gonzalez [3]	1; 32; F	US, CT, MRI, MRCP	left lateral segmen- tectomy	unspecified
Baudin [4]	1; 40; F	US, MRI, MRCP	left hepatectomy	6 months
Van Steenberg [5]	2; 46, 40; F	US, CT, ERCP	left hepatectomy	19–103 months
Nunes [6]	1; 28; F	US, CT, ERCP	left hepatectomy	unspecified
Yu [7]	1; 58; M	US, ERCP, MRI, MRCP	left lobectomy	unspecified
Seidel [8]	1; 43; F	US, ERCP, MRI, MRCP	left lateral segmen- tectomy	unspecified
Delis [9]	1; 50; F	US, CT, ERCP	right hepatectomy	18 months

Table 2. CT findings for differentials of hepatic cystadenoma [adapted from 11, 15]

Tumour	CT finding	Age group
Cystadenoma	septated multi-locular mass; regular with no gross nodules; no intracystic mass	middle-aged women
Cystadenocarcinoma	irregular wall enhancement; septal calcifications; pedunculated outgrowths; intracystic masses	men or women in late life
Simple hepatic cyst	round, regular and unilocular; no enhancement	any, accumulate with age
Hydatid cyst	wall calcifications; hypodense inner matrix; rim enhancement with contrast; multiple in 75%	any
Caroli disease	central dot sign; communicates with biliary lumen	young adults
Undifferentiated sarcoma	septated mass; irregular enhancement	children
Hepatocellular carcinoma	larger solid component; mosaic pattern; venous invasion	usually secondary to risk factors

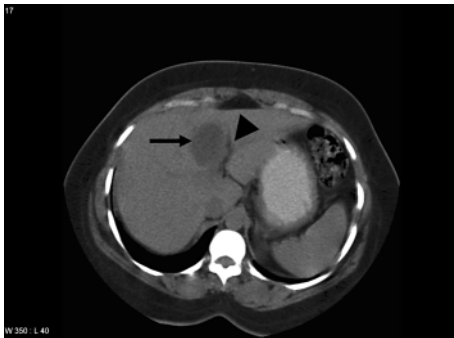
Fig. 1. Representative CT scan image with cyst in liver segment IV (arrow) along with left hepatic duct dilatation (arrowhead).

Fig. 2. MRCP showing cyst (arrow) and compression of the bile duct at the confluence (arrowhead).

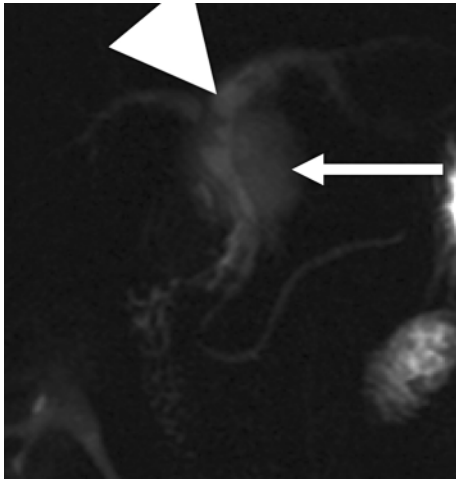
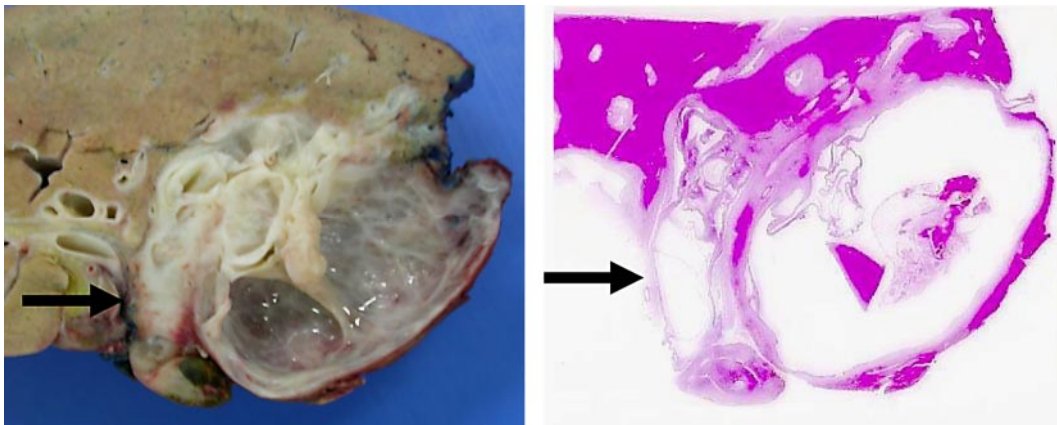


Fig. 3. Gross surgical specimen of left hepatectomy and histology slide showing ball-valve obstruction of the left hepatic duct (arrow) caused by the protrusive element of the biliary cystadenoma.



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