

# Extrahepatic Biliary Cystadenoma with Mesenchymal Stroma: a True Biliary Cystadenoma? A Case Report

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## Abstract

Biliary cystadenomas are benign but potentially malignant cystic neoplasms, which classically contain mesenchymal stroma similar to ovarian tissue. We report a case of an extra-hepatic biliary cystadenoma with mesenchymal stroma along with a discussion of current pathological opinion. **Case presentation:** A 54-year-old female presented with abdominal pain, abnormal liver function tests and a mass on ultrasound. Computerized Tomography identified a complex multi-locular cyst in the common hepatic duct. Radical excision of the lesion and a Roux-en-Y loop bilio-enteric anastomosis was performed. Histology confirmed the presence of a benign biliary cystadenoma with ovarian type stroma. **Conclusion:** Biliary cystadenomas classically contain mesenchymal stroma similar to ovarian tissue. It now appears that cystadenomas without mesenchymal stroma appear to be more akin to similar cystic lesions of the pancreas, and may represent a dissimilar neoplasm. Therefore, malignant transformation can occur, so complete excision is recommended.

## Key words

Cystadenoma – biliary cystic tumour – cysto-adenocarcinoma – mesenchymal stroma – intra-ductal papillary neoplasm – hepatic neoplasm

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## Introduction

Biliary cystadenomas are rare cystic neoplasms, that occur in the intra-hepatic and less commonly in the extra-hepatic ducts [1]. These lesions can present with abdominal pain, distension and nausea. Radiological imaging is

fundamental in diagnosis, and a cystadenoma should be considered whenever an ovoid multi-locular cystic lesion is detected in the hepatic ducts. Assumption that these lesions are benign, based on radiological findings is unadvised and complete excision is recommended [2, 3]. Approximately 80% of biliary cystadenoma contain mesenchymal stroma similar to ovarian tissue; these lesions occur exclusively in females, whereas cystadenomas without mesenchymal stroma occur in both sexes and tend to arise in older patients. This variant is associated with a poorer prognosis and appears to be more closely related to a similar lesion that occurs in the pancreas, termed an intra-ductal papillary neoplasm of the pancreas (IPN-P) [4]. We describe a case of an extra-hepatic biliary cystadenoma with mesenchymal stroma and discuss the current pathological opinion.

## Case presentation

A 54-year-old female presented with abdominal pain, abnormal liver function tests (LFTs) and a biliary mass on ultrasound (US). Clinical examination was normal; a repeat US confirmed the presence of an intra-luminal polypoid multi-locular cystic mass in the common hepatic duct with moderate dilatation of the intra-hepatic ducts. Computerized Tomography (CT) confirmed presence of a complex multi-locular mass in the common hepatic duct (Fig. 1). The patient underwent a laparotomy through a reverse L incision, this approach was favoured because it allows excellent exposure and is associated with reduced morbidity. A distended choledochus was identified and a mass was palpable. The hilar structures were identified and a retrograde dissection of the gallbladder was performed. The common bile duct (CBD) was divided above the duodenum and below the hilum and a Roux-en-Y loop bilio-enteric anastomosis was performed to re-establish biliary drainage. The patient made an excellent recovery and no evidence of recurrence was found on follow up. A 1.8 x 1.5 x 1.4 cm cystic lesion that was removed from the CBD lumen (Fig. 2) was composed of ovarian type stroma containing cysts that were lined by columnar biliary type epithelium (Fig. 3). Focal ulceration and granulation was present but there was

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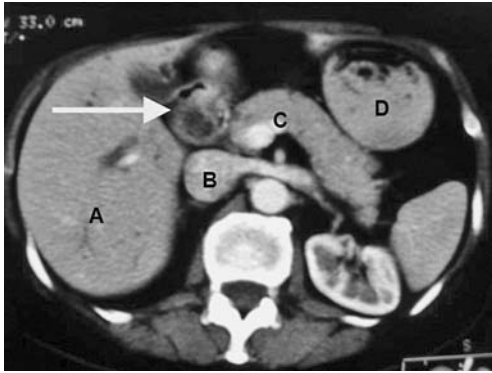
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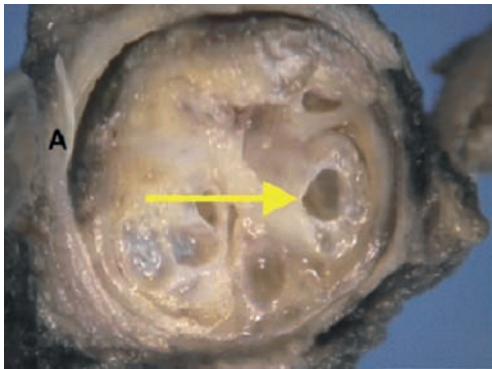
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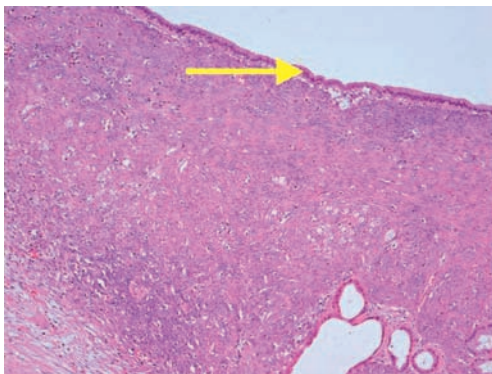
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**Fig 1.** CT abdomen: low-density well lobulated multi-locular thick walled, cystic mass with internal septa, marked with arrow. A = liver, B = pancreas, C = duodenum, D = stomach.



**Fig 2.** Cross section of the common bile duct: a multi-locular cystic lesion occluding the lumen. Arrow demonstrating thick walled internal septations. A = common hepatic duct wall.



**Fig. 3.** Biliary cystadenoma showing ovarian type stroma containing cysts lined by columnar biliary type stroma. Arrow demonstrating columnar epithelium (H&E, 100x).

no dysplasia. Immuno-histochemical staining demonstrated both oestrogen and progesterone receptors (Fig. 4). The conclusion was a benign biliary cystadenoma with ovarian type stroma.

## Discussion

Biliary cystadenomas are rare benign cystic masses with smooth external surfaces, that contain mucinous or serous locules. They occur at any age, but most commonly present in middle aged females (90%) [5]. The clinical presentation depends on tumour location; intra-hepatic tumours are more common, present later, and tend to be larger - 1.5cm to 35cm [5]. Extrahepatic biliary tumours are smaller and present earlier, usually with abdominal pain and obstructive jaundice [6].

Abdominal US demonstrates a thick walled ovoid cystic mass, either anechoic or hypoechoic. Internal septations that make up the multi-locularity are echogenic, while papillary projections could indicate the presence of malignancy [7]. CT reveals a hypodense, well lobulated thick walled cystic mass with internal septa [7]. The mass wall is well defined and enhances with the administration of contrast material. Irregular thickness of the wall, mural nodules or papillary projections again indicate the possibility of malignancy [7]. Magnetic Resonance Imaging (MRI) T1-weighted images show a fluid containing, multi-locular, septated homogeneous low signal intensity mass, Gd-DTPA administration causes enhancement of the walls and septa. T2-weighted images demonstrate fluid collections within the tumour with variable homogeneous high signal intensity while the wall of the mass is represented by a low-signal-intensity rim [8]. So far no radiological imaging can determine the presence of mesenchymal stroma.

Macroscopically, biliary cystadenomas are classically globular or rounded masses with smooth external surfaces from which cysts of various sizes bulge. The inner portion of the cyst is formed of locules; the walls are thin, smooth and glisten. The contents of the locules can be mucinous or serous, or can appear purulent if infected or bloody if there has been a haemorrhage [4]. Microscopically, there is a single layer of epithelial cells that is either lined by biliary-type cuboidal or non-ciliated columnar cells. The majority of cystadenomas have mesenchymal stroma; while this resembles ovarian stroma, it is more akin to the primitive mesenchyme of the embryonic gallbladder and bile ducts [9]. However, this mesenchyme does stain positive for oestrogen and progesterone receptors. Cystadenomas without mesenchymal stroma contain a stroma of hyalinized collagen, and do not stain for oestrogen and progesterone receptors [10].

Comparable lesions are described in the pancreas, where cystic lesions without mesenchymal stroma are termed IPN-P. These are seen as distinctive lesions that differ in presentation, immuno-phenotype, immuno-histochemical expression and outcome to pancreatic cystadenomas. IPN-P appear to be more similar to biliary cystadenoma in presentation and prognosis; furthermore, both lesions contain hyalinized collagen and are immuno-phenotypically and immuno-histochemically similar [2]. In addition, the lack of hormonal receptors suggests an alternative pathogenesis to a cystadenoma with mesenchymal stroma. Altogether, this

suggests that biliary cystadenoma without mesenchymal stroma may represent a cystic variant of intra-ductal papillary neoplasia of the biliary ducts (IPN-B) rather than a true biliary cystadenoma [2, 9]. As such it has been proposed that the term biliary cystadenoma should be reserved for biliary cystic lesions that contain mesenchymal stroma alone [2]. Due to the potential malignancy and the transformation into cystadenocarcinoma, nothing less than complete excision is recommended.

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