

CASE REPORT

Papillary Adenoma of the Distal Common Bile Duct Associated with a Synchronous Carcinoma of the Peri-Ampullary Duodenum

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ABSTRACT

Context Benign tumours of the biliary tract are an extremely rare group of neoplasms. The diagnosis of these rare tumours is established on histopathological analysis following resection. Coincidence of a biliary adenoma of the distal common bile duct and a synchronous adenocarcinoma of the peri-ampullary duodenum has never been reported in the literature.

Case report We report a case of a papillary adenoma in the common bile duct in a 75-year-old female, who had synchronous invasive adenocarcinoma of the peri-ampullary duodenum.

Conclusion Isolated papillary adenoma of the bile duct is extremely rare, and in this unusual case it coincided with a peri-ampullary duodenal adenocarcinoma. However, this is a rare instance of an incidental finding within the distal bile duct following pancreaticoduodenectomy for curative treatment of a peri-ampullary adenocarcinoma.

INTRODUCTION

Adenomas of the extrahepatic bile ducts are benign, polypoid, epithelial neoplasms composed of papillary, tubular, or combined tubulopapillary structures, lined by dysplastic epithelium. In contrast to adenomas of the ampulla, adenomas of the extrahepatic bile

ducts are exceedingly rare and current literature on this topic is sparse. Clinical presentation of adenomas tends to be similar to malignant tumours, hence posing a diagnostic challenge [1]. A few of these rare tumours have been reported in patients with familial adenomatous polyposis [2] but our patient did not exhibit any clinical signs of familial adenomatous polyposis or Gardner's syndrome [3].

We report a case of a 75-year old woman with papillary adenoma of the distal common bile duct and a synchronous adenocarcinoma of the peri-ampullary duodenum.

CASE REPORT

A previously fit 75-year-old lady presented with jaundice, weight loss and abnormal liver function tests. Endoscopic retrograde cholangio-pancreaticography (ERCP) revealed an oedematously enlarged ampulla of Vater and choledocholithiasis, and hence, sphincterotomy and extraction of calculi were performed. The patient re-presented three months later with weight loss, anaemia and abnormal liver function tests. Magnetic resonance cholangio-pancreaticography (MRCP) demonstrated a 13 mm filling defect in the distal bile duct consistent with recurrent calculus. Repeat ERCP demonstrated an abnormal appearing ampulla of Vater raising suspicion of a tumour. Brush cytology was reported as benign. Endoscopic ultrasound scan (EUS) and computed tomography (CT)



Figure 1. Computed tomography (CT) confirmed the presence of a 17 mm intra-luminal soft tissue mass in the peri-ampullary region with no evidence of metastasis.

confirmed the presence of an intraluminal soft tissue mass in the peri-ampullary duodenum with no evidence of metastasis (Figure 1). The patient underwent a pancreaticoduodenectomy with Roux-en-Y reconstruction. She made an uneventful recovery and remains well nine months post-surgery.

Histological analysis revealed moderately and poorly differentiated intestinal-type adenocarcinoma of the peri-ampullary duodenum with continuous infiltration of the ampulla, pancreatic parenchyma, distal main pancreatic duct and distal common bile duct. The invasive adenocarcinoma, which measured approximately 3.6x2.1x3.0 cm (Figure 2a), arose within an extensive, carpet-like tubular adenoma that involved most of the second part of the duodenum, and showed continuous extension into some Brunner's glands and the ampulla (stage pT4). A separate, 0.7x0.4 cm papillary adenoma of biliary type showing low-grade dysplasia was present in the extrapancreatic common bile duct at the junction of the cystic duct (Figures 2b, 3a, 3b). There was no (flat) dysplasia of the bile duct epithelium in between the adenoma and the ampulla of Vater. All 12 lymph nodes identified were tumour free.

DISCUSSION

The World Health Organization (WHO) [4] classification of benign epithelial tumours of the gallbladder and extrahepatic bile ducts

includes tubular, papillary and tubulopapillary adenomas, biliary cystadenoma and papillomatosis (adenomatosis). Prior to this classification, case reports used a variety of terminology to describe bile duct adenomas, including tubular adenoma, villous adenoma, tubulovillous adenoma, adenoma not otherwise specified and papilloma [5].

Traditionally, these adenomas are subdivided into intestinal and biliary types with the latter actually never having been reported in the extrahepatic bile ducts. Pyloric type which is the adenoma found commonly in the gallbladder and frequently associated with stones has also never been reported in this location [5]. Subscribing to this pattern, the bile duct adenoma in our patient was of an intestinal type.

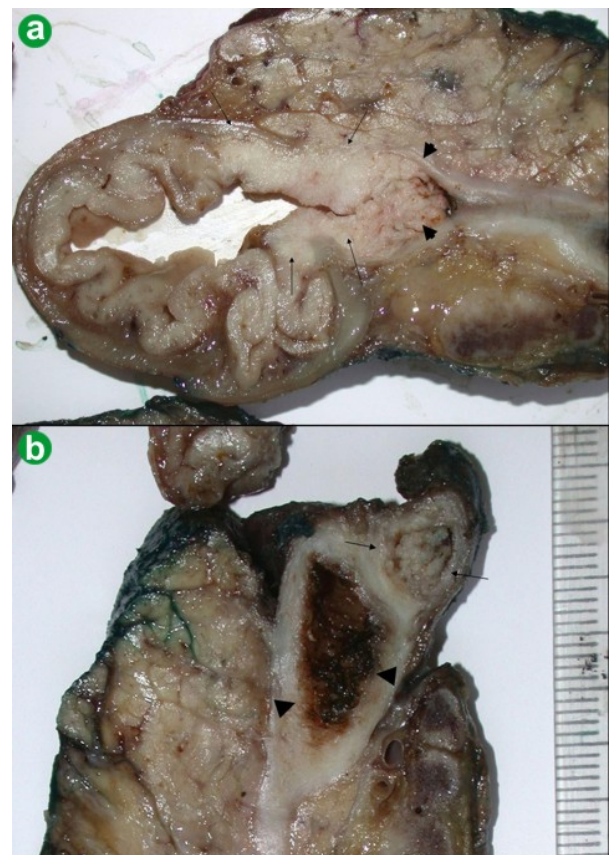


Figure 2. a. A peri-ampullary adenocarcinoma (arrows) involves the duodenal wall and ampulla and extends in a polypous-exophytic fashion (arrow heads) into the distal end of the dilated common bile duct. **b.** The biliary adenoma presents macroscopically as a well-circumscribed exophytic polypous lesion (arrows) in the extrapancreatic portion of the dilated common bile duct (arrow heads).

While adenomas of the gallbladder often remain asymptomatic, adenomas of the extrahepatic biliary tree usually present clinically with recurrent cholangitis, painful jaundice, chronic right upper quadrant pain, or an abdominal mass. In the present case, the adenoma was an incidental finding following treatment for gallstones in the common bile duct and a suspected duodenal tumour. The relationship between adenoma of the extrahepatic bile ducts and choledocholithiasis is not clear. The marked difference in incidence between both pathologies suggests there to be no aetiological association [5]. Although there may be occasional reports on patients with coincidental bile duct adenoma and choledocholithiasis - as in the current case - they do not provide convincing evidence for this association to be more than merely fortuitous. Interestingly, 50-60% of

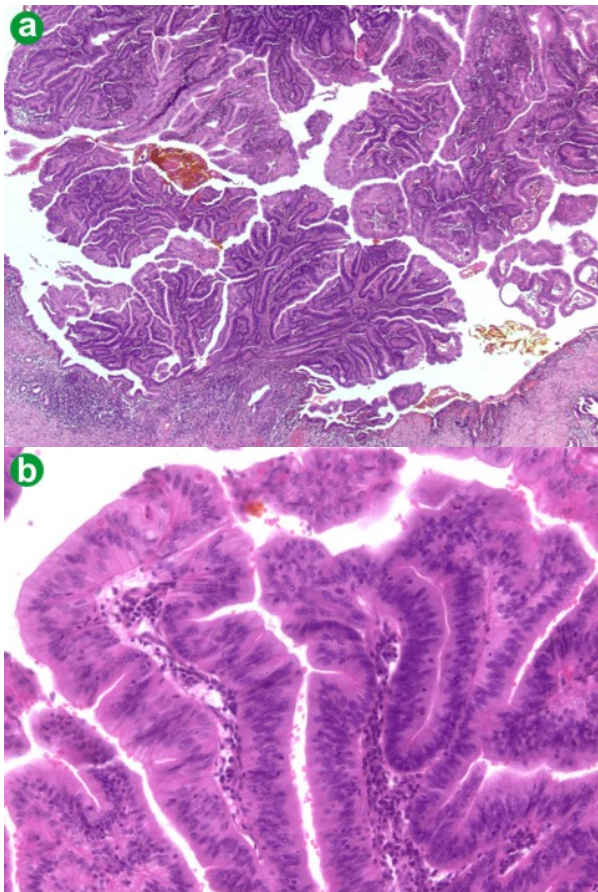


Figure 3. a. The biliary adenoma consists of papillary intraluminal projections lined with dysplastic columnar epithelium (H&E x25). **b.** Neoplastic epithelium lining the papillary projections of the biliary adenoma shows features of low-grade dysplasia (H&E x200).

adenomas arising in the gallbladder are associated with cholelithiasis, however, these adenomas are often of a pyloric gland type, which has never been reported in the extrahepatic bile ducts. These differences in histology and association with lithiasis underscore the disparate biology of adenomatous neoplasia in the gallbladder and distal bile duct.

In the absence of gallstones, these extrahepatic bile duct adenomas are thought to remain dormant for long periods (up to 14 years) [6] and they may eventually cause vague symptoms due to local pressure effects [7]. Pre-operative differentiation between papillary adenomas and malignant tumours is extremely difficult due to the rarity of occurrence, similar clinical presentation and lack of definitive diagnosis based on radiological imaging alone.

Imaging modalities, including CT, MRCP, ERCP and EUS, have been used in identifying these tumours with limited success. Stewart *et al.* demonstrated that cytological analysis alone could not differentiate bile duct adenomas from adenocarcinoma in three patients in a study of 406 patients with pancreatobiliary strictures [8]. Hence, pre-operative diagnosis of extrahepatic bile duct adenomas is extremely difficult.

Our case report illustrates synchronous occurrence of an exceedingly rare tumour i.e. bile duct adenoma and a duodenal adenocarcinoma with precursor adenoma. The clinical importance of the bile duct adenoma has to be considered in the context of only a small number of them progressing to carcinoma. This view is further substantiated by the fact that adenomas are much less common than carcinomas of the extrahepatic bile ducts, and that most carcinomas do not show evidence of a residual adenoma. In fact the mucosa adjacent to the carcinomas not uncommonly displays flat dysplasia and carcinoma in situ, which are believed to be the carcinoma precursor [5].

Due to the limited understanding of their malignant potential, there are no guidelines for the management of papillary adenomas. In

almost all cases, since malignancy is suspected pre-operatively, surgery with intent to cure should be performed when feasible. In the present case, a pancreaticoduodenectomy was performed to treat a peri-ampullary adenocarcinoma, and the papillary adenoma was an incidental finding within the distal common bile duct.

CONCLUSION

Papillary adenoma of the extra pancreatic bile duct is a rare entity and it should be borne in mind as a differential for all patients with obstructive jaundice. By multimodal imaging and with the aid of transpapillary biopsy or brush cytology a working diagnosis may be reached. Frequently, ERCP and CT scanning will give the additional information before surgery is embarked upon. The surgical procedure of choice in cases of suspected malignancy remains pancreaticoduodenectomy.

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