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Radiologic-Pathologic Correlations

Intraductal growth–type mucin-producing peripheral cholangiocarcinoma associated with biliary papillomatosis

Mine G. Güllüoglu, MD[a,*], Ilgin Ozden, MD[b], Arzu Poyanli, MD[c], Ugur Cevikbas, MD[a], Orhan Ariogul, MD[b]

[a]*Department of Pathology, Istanbul Faculty of Medicine, Istanbul University, 34390 Istanbul, Turkey*
[b]*Department of General Surgery, Istanbul Faculty of Medicine, Istanbul University, 34390 Istanbul, Turkey*
[c]*Department of Radiology, Istanbul Faculty of Medicine, Istanbul University, 34390 Istanbul, Turkey*

Abstract A 64-year-old woman with upper abdominal pain, nausea, and vomiting was admitted. The magnetic resonance imaging revealed marked dilation and "crowding" of the segment 4 bile ducts with an area suspicious for a stone or tumor. Ultrasonography-guided percutaneous transhepatic cholangiography revealed multiple filling defects in the segment 4 bile ducts, the left and common hepatic ducts. A left hepatectomy and cholecystectomy was performed. Dilated bile ducts containing mucinous material and a mass in the cystically dilated bile ducts of segment 4 were detected in the gross examination. It showed continuity within the surrounding dilated bile ducts. The dilated bile ducts of the segments 2 and 3 contained mucinous material without any apparent mass formation. Microscopically, the bile ducts were lined by biliary epithelium displaying simple and complex papillary structures with moderate to severe degree of dysplastic changes. The mass was composed of complex papillary structures filling the bile duct with a few foci of invasion. The papillary structures were composed of mucin-producing columnar cells as well as cells with oncocytic appearence. Patchy cytokeratin 7, cytokeratin 19, hepatocyte paraffin 1, MUC2, and CDX2 immunopositivities were observed. Biliary papillomatosis, mucin-producing intrahepatic cholangiocarcinoma, and intraductal papillary-type peripheral cholangiocarcinoma are in the same disease spectrum of papillary biliary neoplasm and termed as *intraductal papillary neoplasm of the liver*. Mucinous hypersecretion and signs of mucobilia are considered specific and should raise the suspicion of lesions in this spectrum.

Keywords: Biliary papillomatosis; Cholangiocarcinoma; Intraductal papillary neoplasm; Mucobilia; Percutaneous transhepatic biliary drainage

1. Introduction

Biliary papillomatosis (BP) is a rare entity which has been recognized more frequently with developing radioimaging techniques [1-4]. It is characterized by multiple mucosal papillomatous lesions at various stages in the adenoma-carcinoma sequence in the biliary tree [5]. The involvement may be intrahepatic, extrahepatic, or diffuse [2]. Mucin-producing and non–mucin-producing variants have been reported [1]. Although it was initially considered to be a relatively benign lesion, accumulating experience reflects a significant risk of malignant transformation [1,4].

2. Case report

A 64-year-old woman who had been under treatment for chronic obstructive pulmonary disease and anxiety disorder, applied with complaints of upper abdominal pain, nausea, and vomiting. She had previously applied to another institution with the same complaints 9 months ago, and the radiologic evaluations had revealed dilated bile ducts of the left lobe of the liver, and no obvious cause of biliary obstruction had been found. Blood biochemistry including liver function tests were within normal limits. The findings had been interpreted as Caroli's disease isolated to the left liver lobe, and conservative follow-up had been recommended. The biochemical findings as well as radiologic findings were unchanged after a follow-up period of six months.

* Corresponding author. Tel.: +90 212 414 23 98; fax: +90 212 631 13 67.
E-mail address: mggullu@istanbul.edu.tr (M.G. Güllüoglu).

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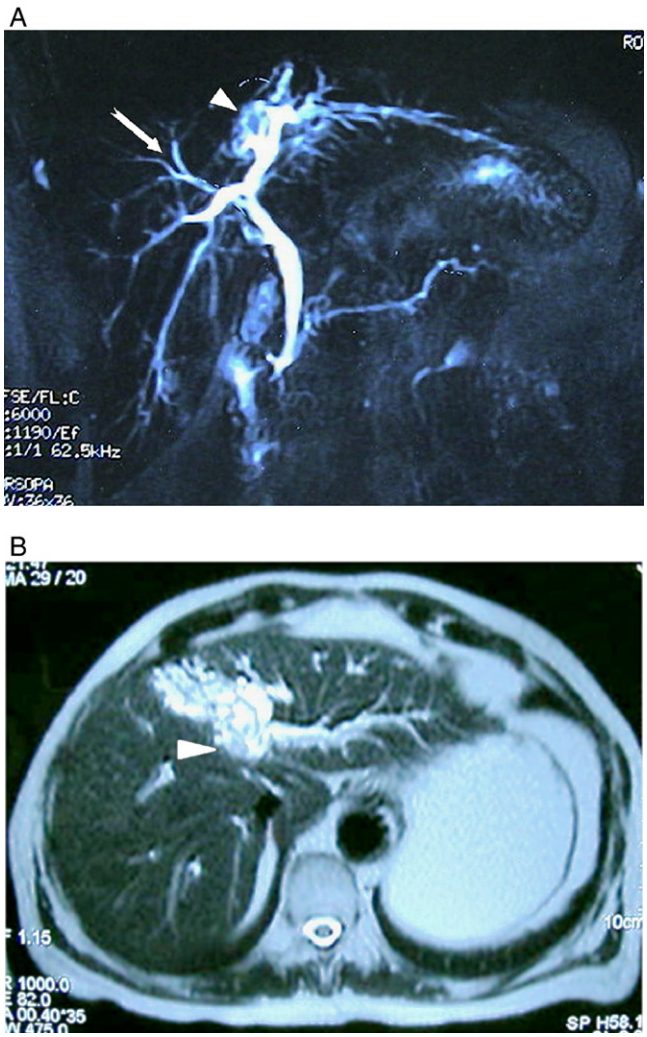

Fig. 1. (A) The bile duct of the right posterior section joins the left hepatic duct both of which are dilated in comparison with the right anterior section duct (arrow). The tumor is in segment 4 (arrowhead). (B) The axial section shows dilation of the biliary tree in the left lobe and the tumor in segment 4 (arrowhead).

Physical examination was unremarkable except for a pain upon palpation in the right upper abdominal quadrant and the epigastrium. Levels of aspartate aminotransferase, alanine aminotransferase, alkaline phosphatase, gamma-glutamyl transpeptidase, total and direct bilirubin, as well as carcinoembryonic antigen (CEA) and carbohydrate antigen 19-9 were within normal limits. Upper gastrointestinal endoscopy and endoscopic biopsy revealed hiatal hernia and chronic *Helicobacter pylori* gastritis, respectively. The magnetic resonance imaging revealed marked dilation and "crowding" of the bile ducts of the segment 4 (Fig. 1). The small signal-void area in segment 4 was considered suspicious for a stone or tumor. The bile duct of the right posterior section joined the left hepatic duct; the bile ducts of the left lateral section and the right posterior section were mildly dilated. However, no mass lesion that caused obstruction was observed. Ultrasonography-guided percutaneous transhepatic cholangiography was performed.

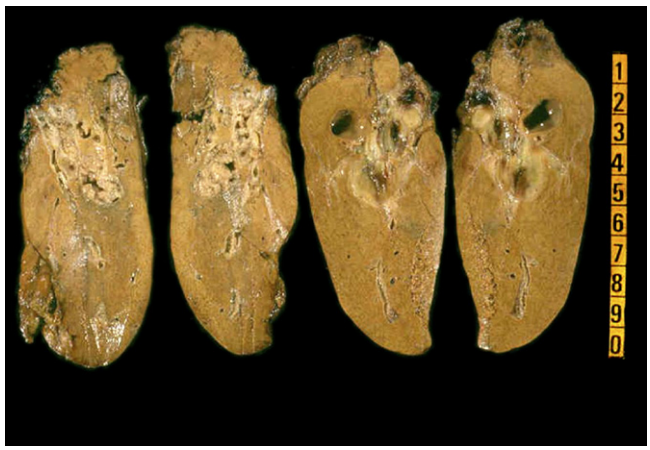

Fig. 2. The mass lesion in the dilated and mucin-filled bile ducts.

There were multiple filling defects in the segment 4 bile ducts as well as in the left hepatic duct and the common hepatic duct. However, contrast flowed freely into the right

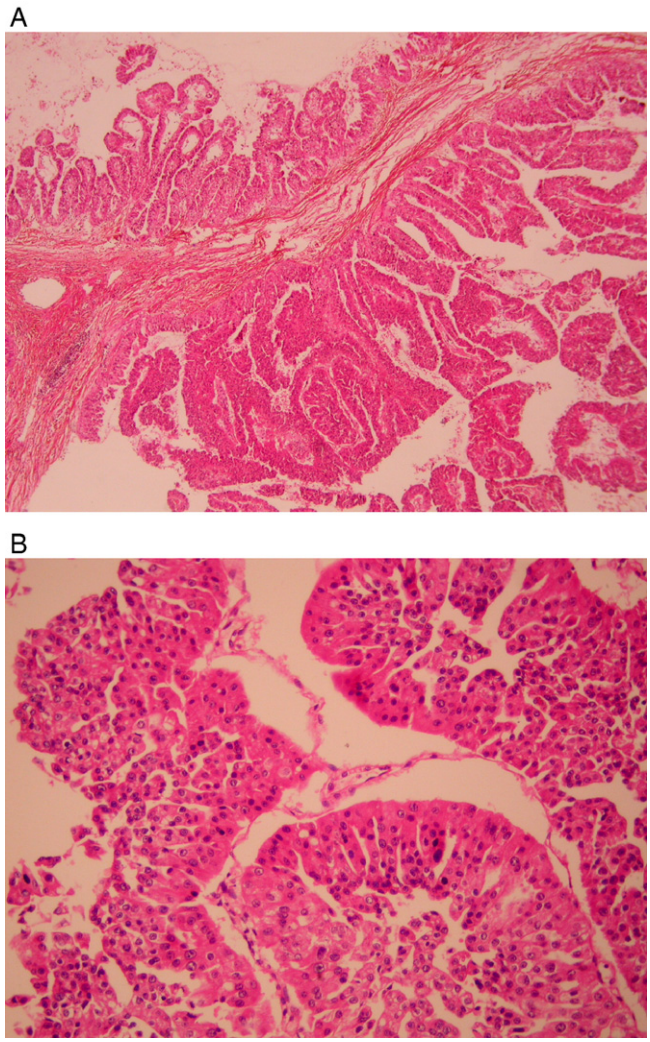

Fig. 3. (A) Papillary structures composed of cells with large acidophilic granular cytoplasm (B) and cells with periodic acid-Schiff (PAS) (+) clear cytoplasm.