



# Heterotopic chondroid tissue of the main bile duct mimicking Klatskin tumor: case report and review of the literature

Fatma Hüsniye Dilek<sup>1</sup> · Şebnem Karasu<sup>2</sup> · Osman Nuri Dilek<sup>3</sup>

Received: 13 October 2018 / Accepted: 22 November 2018 / Published online: 29 November 2018  
© Japanese Society of Gastroenterology 2018

## Abstract

Heterotopic tissue in the bile duct is a very rare condition. There are a few case reports of heterotopic tissue including gastric and pancreatic cells. However, we could not find any data regarding heterotopic chondroid tissue obstructing the common bile duct in the literature. A 56-year-old woman was admitted to our hospital with complaints of progressive jaundice and pruritus. Laboratory test results were within the normal limits except bilirubin and alkaline phosphatase which were 10.8 mg/dL and 256 IU/L, respectively. Endoscopic retrograde cholangiography (ERCP) and computed tomography (CT) revealed a biliary stricture confined to the upper part of the common bile duct. The patient was operated as soon as the bilirubin level dropped below 5 mg/dL by percutaneous transhepatic cholangiography. Despite the lack of a precise confirmation, the patient was prepared for operation considering the possibility of a malignant biliary tumor since the radiographic mass findings together with hyperbilirubinemia were highly suggestive of malignancy. The patient underwent total extrahepatic bile duct resection and Roux-en-Y hepaticojejunostomy, and was discharged without any postoperative complications. Histologic examination of the bile duct and the nodule revealed the presence of a well-demarcated chondroid tissue within the subepithelial connective tissue. Herein, we presented the first case, to our knowledge, of heterotopic chondroid tissue of the common bile duct in the literature. It is a benign condition that should be considered in the differential diagnosis of stricture and mass-forming lesions of the bile duct.

**Keywords** Bile duct · Chondroma · Heterotopic tissue · Jaundice · Klatskin tumor

## Introduction

Obstructive jaundice in the adult usually occurs as a consequence of periampullary tumors and bile stones. Less frequently, benign tumors or tumor-like lesions might also lead to obstructive jaundice mimicking malignancies; that

could end up with major operations such as hepatectomy and pancreatoduodenectomy [1].

Chondroid heterotopia within the common bile duct has not been previously reported in the English literature [2–4]. Herein, we present a case that has been treated with biliary tract resection due to a biliary tract chondroma.

✉ Osman Nuri Dilek  
osmannuridilek@gmail.com

Fatma Hüsniye Dilek  
fhdilek@hotmail.com

Şebnem Karasu  
sebnem.karasu@gmail.com

<sup>1</sup> Department of Pathology, School of Medicine, İzmir Katip Çelebi University, İzmir, Turkey

<sup>2</sup> Department of Radiology, Atatürk Education and Research Hospital, İzmir Katip Çelebi University, İzmir, Turkey

<sup>3</sup> Department of Surgery, School of Medicine, İzmir Katip Çelebi University, İzmir, Turkey

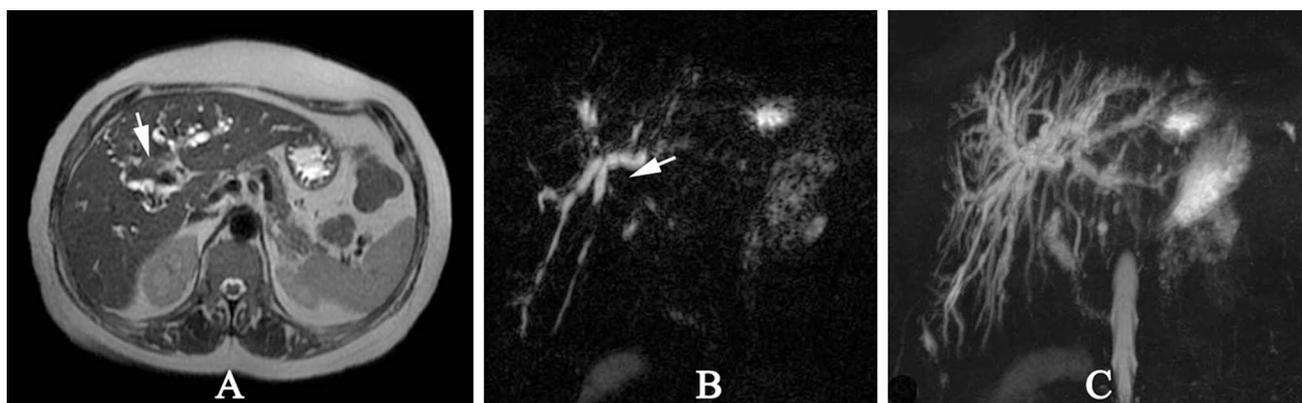
## Case

A 56-year-old woman was admitted to our hospital with complaints of progressive jaundice and pruritus. Laboratory test results were as follows: total bilirubin 10.8 mg/dL, alkaline phosphatase 256 IU/L, amylase 56 U/L, and C-reactive protein 3.6 mg/dL. Tumor markers including CA19-9 and CEA were within the normal limits. Endoscopic ultrasonography (EUS) could not detect the lesion, therefore, fine needle aspiration biopsy (FNA) was not performed. A nodular soft tissue lesion in the liver hilum and obstructive dilatation in the intrahepatic bile ducts

were detected on contrast-enhanced CT (Fig. 1a–c). ERCP showed a biliary stricture confined to the upper part of the common bile duct. Following an unsuccessful ERCP intervention, the patient was operated by percutaneous transhepatic cholangiography (PTC) as soon as the bilirubin level dropped below 5 mg/dL. Despite the lack of a precise confirmation, the patient was prepared for operation considering the possibility of a malignant biliary tumor since the radiographic mass findings together with hyperbilirubinemia were highly suggestive of malignancy. Informed consent form has been signed by the patient as part of the routine procedure. Following the preoperative preparation, laparotomy has been performed for definitive diagnosis and treatment. The lesion was located just above the entrance of the cystic duct into the common bile duct. The distance between hepatic duct confluence and the cystic duct conjunction was measured as 2 cm (Fig. 2a). Radiologic findings were in favor of a Klatskin tumor type 1 or

2 according to Bismuth–Corlette classification. The patient underwent total extrahepatic bile duct resection and Roux-Y hepaticojejunostomy. Histopathologic evaluation of surgical margins via frozen section biopsies was normal. Lymphadenopathy was not present. The patient recovered well after the surgery and has been asymptomatic for the last 10 months.

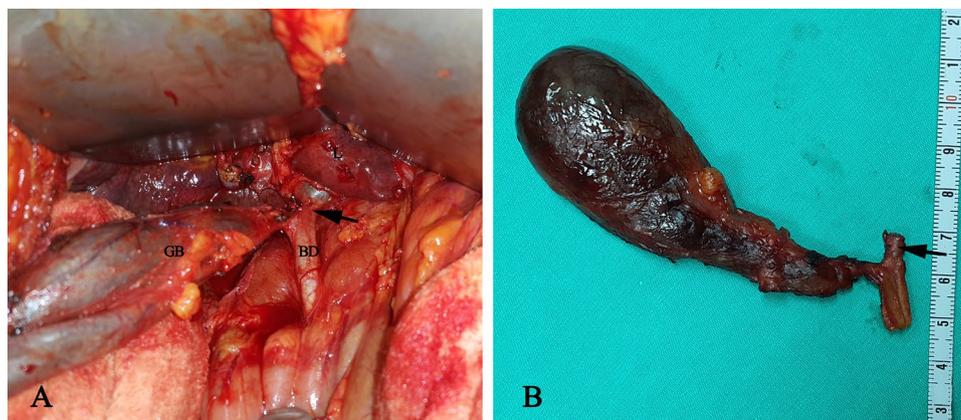
Macroscopic examination revealed an obstructing submucosal nodule located in the upper part of the common bile duct, just above the level of ductus cysticus (Fig. 2b). The nodule measured 0.7 mm in diameter with a homogenous, white, and glistening cut surface (Fig. 3a). Histologic examination of the bile duct and the nodule revealed a well-demarcated chondroid tissue within the subepithelial connective tissue (Fig. 3b). Chondrocytes were mature and lacked significant cellular and nuclear atypia. Additionally, mononuclear cells consistent with a minimal chronic inflammatory process in the bile duct wall were observed.



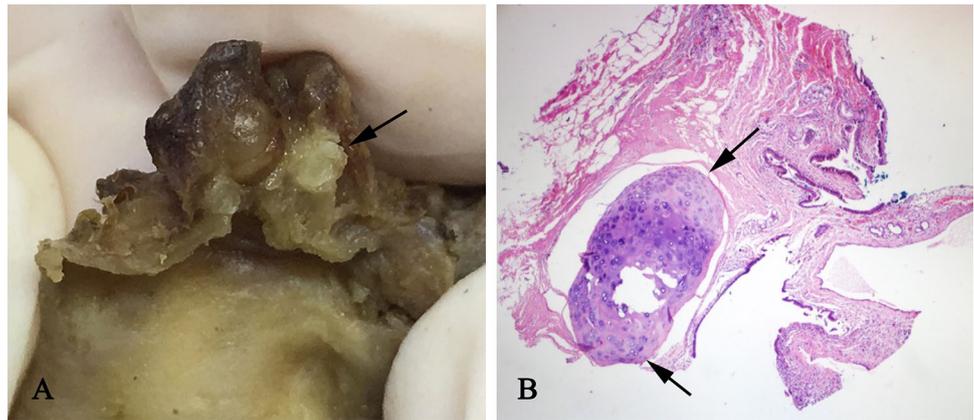
**Fig. 1** A contrast-enhanced computed tomography shows on T2W transverse MR image (a), a nodular soft tissue lesion is seen in the liver hilum. The lumen signal of the common bile duct cannot be

identified on the source image of MRCP (b) which is compatible with severe stenosis. On MIP image (c) obstructive dilatation is seen in the intrahepatic bile ducts

**Fig. 2** a Biliary stricture just above the cystic duct (arrow) and dilatation of the biliary confluence. Specimen b shows the stricture (arrow). GB gallbladder, BD common bile duct, L liver



**Fig. 3** Macroscopic appearance (a) of chondroid nodule (arrow); homogeneous, white, and glistening and measured 0.7 mm in diameter. Microscopic appearance (b) of well-demarcated heterotopic chondroid tissue (arrow) within the subepithelial connective tissue



## Discussion

Obstructive jaundice is usually associated with malignancies of the pancreatobiliary system in adults. Benign or inflammatory lesions can occasionally mimic malignancy. Therefore, accurate preoperative diagnosis of biliary tract lesions with endoscopic and radiologic imaging might be challenging. EUS could not detect the lesion probably because the lesion is too small and located in the upper common bile duct. However, it is well known that EUS-FNA may have a significant role in the preoperative diagnosis in this kind of bile duct lesions [3]. In cases mimicking malignancy similar to ours, hepatectomy and pancreaticoduodenectomy may be mistakenly performed. In a series of 275 cases thought preoperatively to have malignant neoplasms, Corvera et al. reported that 22 of them were diagnosed as benign lesions histologically [1]. Recently, cases of benign stenosis that mimics malignancy have been described [5–7]. They are included as heterotopic pancreas, gastric tissues and also localized chronic cholangitis. Moreover, Fukuda et al. have reported a case of heterotopic gastric mucosa mimicking hilar cholangiosarcoma [6]. There are several reports of heterotopic chondroid tissues found in the head and neck, uterus and peritoneal cavity [8, 9]. However, to our knowledge, heterotopic chondroid tissue that mimics malignancy has not yet been reported in the English literature.

Some theories have been proposed for the genesis of chondroid heterotopy. Proliferation of immature chondroid cells resting in the head and neck region was held responsible from such pathology. Regarding her age, the present case would make little sense from an embryological perspective. Nonetheless, it suggests that chondroid heterotopia may appear to be a developmental anomaly. Any anomalies during embryological development might lead to the formation of aberrant mesenchymal tissue within various sites. Some authors proposed that proliferation of cartilage in soft tissue probably reflects the multipotential nature of primitive mesenchymal cells, whose growth may be stimulated by trauma,

irritation, or inflammation [7–9]. Fadare et al. have reported two cases of cartilaginous differentiation in peritoneal tissue, that may represent metaplastic lesions of the secondary müllerian system or a unique peritoneal response to previous surgical manipulation [9]. Spanta et al. have suggested mesenchyme of the myosalpinx or subcoelomic mesenchyme of the tubal serosa as the origin of tubal chondroma [10]. Similarly, multipotential cells in the subepithelial mesenchyme of the bile duct might be the origin of chondroid metaplasia in our case. The present case is a unique example of subepithelial mesenchymal differentiation into the chondroid tissue by not being associated to adverse environments such as history of abdominal surgery, trauma or chronic inflammation.

In conclusion, we presented the first case, to our knowledge, of heterotopic chondroid tissue of the common bile duct in the literature. It is a benign condition that should be considered in the differential diagnosis of stenosis and mass-forming lesions of the bile duct.

## Compliance with ethical standards

**Conflict of interest** All authors declare that they have no conflict of interest.

**Human rights** All procedures followed have been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

**Informed consent** This is a case report. As a routine procedure of our clinic, informed consent has been taken for all condition including treatment modalities and also publication.

## References

1. Corvera CU, Blumgart LH, Darvishian F, et al. Clinical and pathologic features of proximal biliary strictures masquerading as hilar cholangiocarcinoma. *J Am Coll Surg*. 2005;201:862–9.
2. Maeda S, Yamanaka N, Tanaka T, et al. Idiopathic benign biliary stricture. *J Hepatobiliary Pancreat Surg*. 1998;5:463–6.

3. Molinari M, Ong A, Farolan MJ, et al. Pancreatic heterotopia and other uncommon causes of non-malignant biliary obstruction. *Surg Oncol.* 2000;9:135–42.
4. Franceschi T, Allias F, Mauduit C, et al. Chondroid nodule in the female peritoneum arises from normal tissue and not from teratoma or conception product. *Virchows Arch.* 2018;473:115–9.
5. Sumiyoshi T, Shima Y, Okabayashi T, et al. Heterotopic pancreas in the common bile duct, with a review of the literature. *Intern Med.* 2014;53:2679–82.
6. Fukuda S, Mukai S, Shimizu S, et al. Heterotopic gastric mucosa in the hilar bile duct mimicking hilar cholangiocarcinoma: report of a case. *Surg Today.* 2013;43:91–5.
7. Aoki T, Kubota K, Oka T, et al. Follicular cholangitis: another cause of benign biliary stricture. *Hepatogastroenterology.* 2003;50:639–42.
8. Erdogan D, Kloek JJ, ten Kate FJ, et al. Immunoglobulin G4-related sclerosing cholangitis in patients resected for presumed malignant bile duct strictures. *Br J Surg.* 2008;95:727–34.
9. Fadare O, Bifulco C, Carter D, et al. Cartilaginous differentiation in peritoneal tissues: a report of two cases and review of the literature. *Mod Pathol.* 2002;15:777–80.
10. Spanta R, Lawrence WD. Soft tissue chondroma of the fallopian tube. Differential diagnosis and histogenetic considerations. *Pathol Res Pract.* 1995;191:174–8.