Carolí’s disease associated with biliary papillomatosis and cholangiolithiasis

A 52-year-old man presented with constant right upper quadrant pain and repeated episodes of fever for 6 months, accompanied with cutaneous and scleral icterus. Laboratory tests suggested a rise in bilirubin. A computed tomography (CT) scan showed obvious dilatation of the intra- and extrahepatic bile ducts, and patchy high density shadows in the intrahepatic bile ducts that were thought to be stones (▶Fig. 1).

Duodenoscopy showed that the orifice of the duodenal papilla was shaped like the mouth of a fish and a large amount of mucus was expelled (▶Fig. 2). T-tube cholangiography confirmed the dilated hepatic bile ducts (▶Fig. 3). The patient then underwent biliary exploration. During surgery, a large amount of mucus could be seen in the bile duct, and the T-tube was indwelling in the common bile duct. After the surgery, the patient still had abdominal pain, the T-tube drainage was problematic with mucus discharge observed, and there was no significant regression of the jaundice.

The patient underwent cholangioscopy 6 weeks after surgery, which showed diffuse and labyrinth-like dilatation of the intrahepatic bile ducts (▶Fig. 4a), which was thought to be Carolí’s disease with cholangiolithiasis. Diffuse papillary protrusions could be seen on the intrahepatic bile duct wall, with a large amount of mucus (▶Fig. 4b). A biopsy was taken, which subsequently confirmed the diagnosis of biliary papillomatosis (▶Fig. 5). Most of the stones were removed under cholangioscopy (▶Video 1). After stone extraction, the patient’s abdominal pain was relieved and the degree of jaundice gradually decreased.

Carolí’s disease is a rare autosomal recessive disorder, which leads to intrahepatic stones and recurrent cholangitis when progressive. It is characterized by a biliary abnormality consisting of segmental or saccular dilatations of the large intrahepatic bile ducts [1, 2]. Biliary papillomatosis is also a rare disease entity with a strong malignant potential, which is characterized by multiple papillary adenomas involving both the intrahepatic and extrahepatic biliary tree [3]. This patient had a wide range of lesions due to papillomas, which could not be completely resected. The only complete and effective treatment for this condition is liver transplantation [4].
Competing interests

None

The authors

Chi Yuan*, Jianrong Liu*, Xianghong Zhou, Chuncheng Wu
Department of Gastroenterology, West China Hospital, Sichuan University, Chengdu, China

*Chi Yuan and Jianrong Liu are considered co-first authors of this work.

Corresponding author

Chuncheng Wu, MD
Department of Gastroenterology, West China Hospital, Sichuan University, No. 37 Guo Xue Alley, Chengdu 610041, Sichuan Province, China
hxwucc@163.com

Video 1 Cholangioscopy showing diffuse dilatation of the intrahepatic bile ducts, with a large amount of mucus and cholangiolithiasis. A biopsy is taken and stones are extracted.

Fig. 4 Cholangioscopic views showing: a diffuse and labyrinth-like dilatation of the intrahepatic bile duct; b a large amount of mucus and diffuse papillary protrusions within the intrahepatic bile ducts.

Fig. 5 Histology of the biopsy specimen showing biliary papillomatosis.
References


Bibliography

DOI https://doi.org/10.1055/a-0640-2859
Published online: 3.7.2018
Endoscopy 2018; 50: E276–E278
© Georg Thieme Verlag KG
Stuttgart · New York
ISSN 0013-726X

ENDOSCOPY E-VIDEOS
https://eref.thieme.de/e-videos

Endoscopy E-Videos is a free access online section, reporting on interesting cases and new techniques in gastroenterological endoscopy. All papers include a high quality video and all contributions are freely accessible online.

This section has its own submission website at https://mc.manuscriptcentral.com/e-videos