Thieme

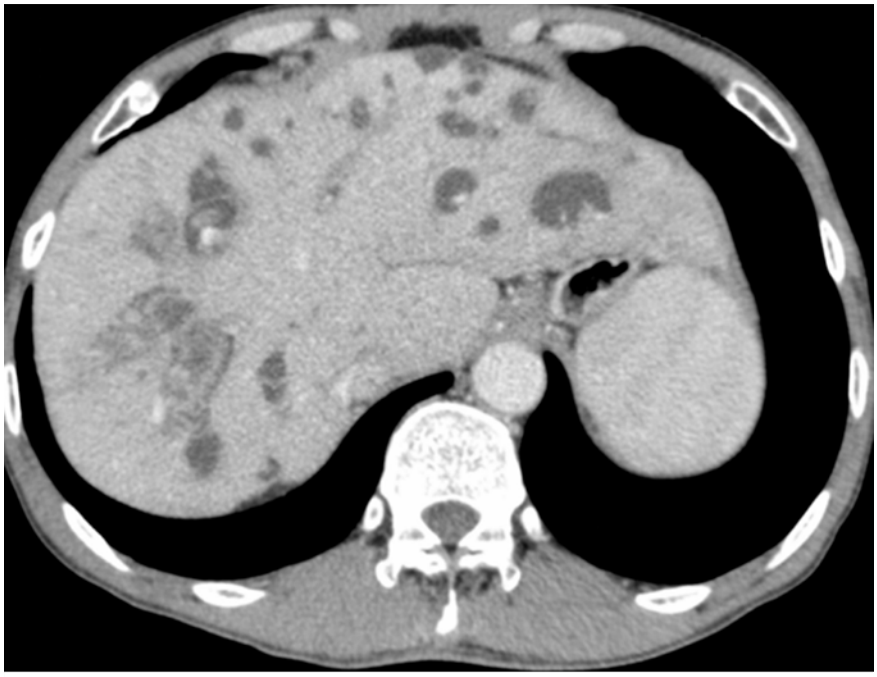
Caroli's disease associated with biliary papillomatosis and cholangiolithiasis

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**).

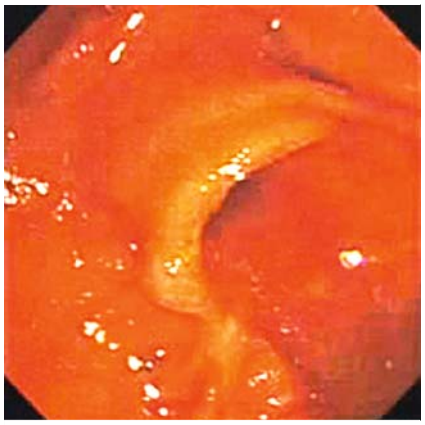
Duodenoscopy showed that the orifice of the duodenal papilla was shaped like the mouth of a fish and a large amount of mucus was expelled (▶ **Fig. 2**). T-tube cholangiography confirmed the dilated hepatic bile ducts (▶ **Fig. 3**). The patient then underwent biliary exploration. During surgery, a large 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 Caroli'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.

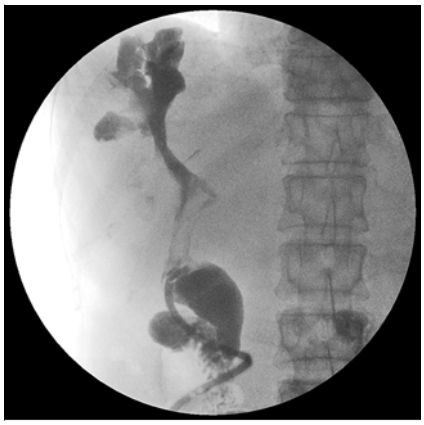
Caroli's disease is a rare autosomal recessive disorder, which leads to intrahepatic stones and recurrent cholangitis when progressive. It is characterized by a biliary abnormality consisting of segmental saccular dilatations of the large intrahepatic bile ducts [1,2]. Biliary papillomatosis is also 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].



▶ **Fig. 1** Computed tomography scan showing obvious dilatation in the intra- and extrahepatic bile ducts, along with patchy high density shadows in the intrahepatic bile ducts that were thought to be stones.



▶ **Fig. 2** Duodenoscopy image showing the orifice of the duodenal papilla, which was shaped like the mouth of a fish.



▶ **Fig. 3** T-tube cholangiogram showing dilatation of the intra- and extrahepatic bile ducts.

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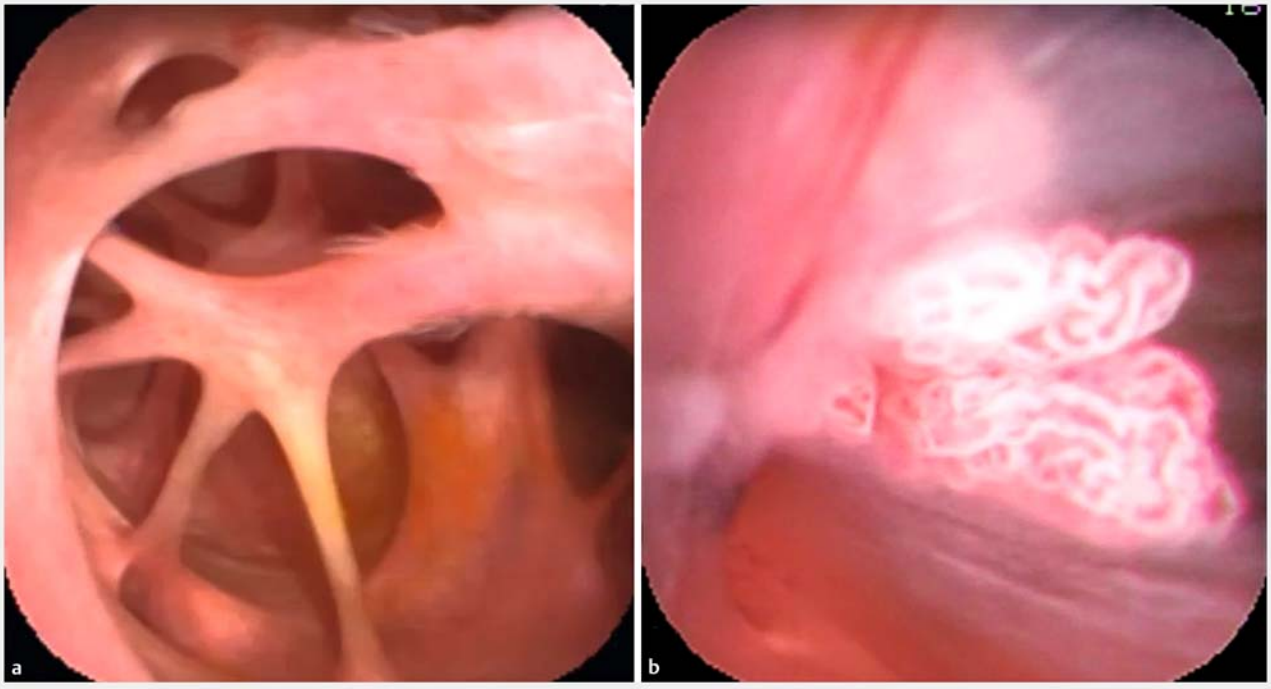
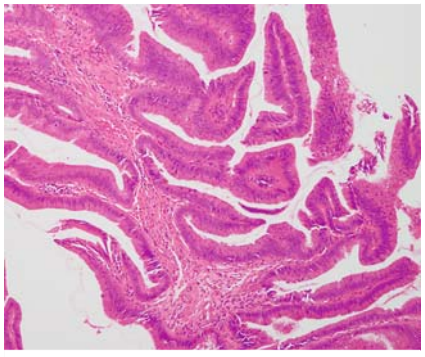
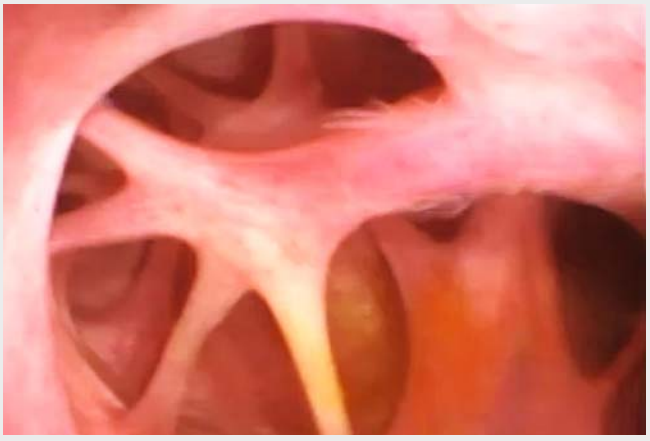


▶ **Fig. 4** Cholangioscopic views showing: **a** diffuse and labyrinth-like dilatation of the intrahepatic bile duct; **b** a large amount of mucus and diffuse papillary protrusions within the intrahepatic bile ducts.



▶ **Fig. 5** Histology of the biopsy specimen showing biliary papillomatosis.



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.

Competing interests

None

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Bibliography
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