Bile duct cysts are defined as dilatations of the extrahepatic or intrahepatic biliary tree. Biliary cysts are classified according to the Todani classification, which encompasses five types. They account for approximately 1% of all benign biliary disease. The incidence of biliary cysts in Europe is 1–2 per 1000000. The Todani type-III cyst is rare and accounts for 2%–4% of all Todani cysts. The risk of developing a cholangiocarcinoma ranges from 2.5% to 28%, and increases with age. Choledocholithiasis, cholelithiasis, hepatolithiasis, cholangitis, pancreatitis, and spontaneous cyst perforation have all been reported in these patients [1,2].

We report the case of a 17-year-old Latino man with systemic candidosis occurring after acute necrotizing pancreatitis (ANP) that was caused by a choledochal cyst. The patient was referred to our unit 35 days after two failed attempts at endoscopic retrograde cholangiopancreatography (ERCP). The patient also had right kidney agenesis. On arrival, the patient’s clinical condition was critical owing to sepsis and cholestasis. A computed tomography (CT) scan was performed, which showed the presence of multiple stones inside the choledochal cyst with a dilated common bile duct (CBD). The pancreatic isthmus and tail contained necrotic areas. An endoscopic ultrasound (EUS) was performed, which showed that the cyst was completely full of stones and occluding the common bile duct.

Using a duodenoscope, we performed a cystotomy, opening the cyst with a needle knife. The stones were then totally removed (Video 1). The patient developed bleeding from the cystotomy 3 days later, which was successfully managed using endoscopic hemostasis. Although the bleeding was controlled, he died the following day from multiorgan failure and systemic sepsis. Acute necrotizing pancreatitis is a life-threatening condition, especially for patients in a poor clinical condition. Endoscopic cystotomy is a possible treatment for choledochal Todani type-III cysts, especially in patients who have undergone an unsuccessful attempt at ERCP. This treatment has not been previously reported.

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Benedetto Mangiavillano, Andrea Parodi, Massimo Conio
Gastroenterology & Gastrointestinal Endoscopy, General Hospital, Sanremo, Italy

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Corresponding author
Benedetto Mangiavillano, MD
Gastroenterology Gastrointestinal Endoscopy
Borea Hospital – Sanremo
Via G. Borea 56
18038 Sanremo
Italy
Fax: +39-0184-536875
b_mangiavillano@hotmail.com

Video 1

Endoscopic ultrasound (EUS) and endoscopic views of the choledochal cyst. Cystotomy is performed with a needle knife, revealing evidence of multiple stones in the opened cyst.