



Direct visualization of biliary stump polyp in a boy with recurrent pancreatitis after surgery for pancreatobiliary maljunction associated with choledochal cyst

Valerio Balassone, MD, PhD,¹ Chiara Imondi, MD,² Tamara Caldaro, MD,¹ Paola De Angelis, MD,¹ Luigi Dall'Oglio, MD¹

BACKGROUND AND AIMS

Pancreaticobiliary maljunctions (PBMs) are congenital anomalies of the pancreatic and biliary duct joint before the duodenal wall and are suspected to be the cause of pancreaticobiliary reflux and biliary dilatation, including congenital choledochal cyst.¹⁻³ Todani's type I cysts are the most frequently associated with PBM (92% of cases).⁴ Untreated PBMs in adults may result in cholangiocarcinoma in 11.1% of cases.^{5,6} PBMs with a long common channel, especially when associated with a choledochal cyst, have an increased incidence of acute pancreatitis (18%-23%).⁷

Pancreatic juice mixed with bile stasis in the dilated common channel may account for the formation of pancreatic stones and protein plugs, causing temporary obstruction of the common channel and bile backflow into the pancreatic duct. The increasing pressure in the pancreatic duct could precipitate acute pancreatitis.^{8,9}

Long-term surveillance is mandatory for patients with PBM because PBM appears to result in a higher risk of biliary cancer, especially in Todani's type I biliary common duct.⁵ The literature reports a biliary cancer risk ranging from 8% to 42% in patients with PBM; in particular, in the Japanese nationwide survey, biliary cancer developed in 22% of adult patients with PBM and biliary dilation and in 42% of adult patients with PBM without biliary dilation.^{2,5}

A high level of suspicion is required for the diagnosis, and the role of ERCP is crucial¹⁰ because MRCP and CT have a limited role in young children. ERCP provides high-sensitivity images, with selective cannulation and the possibility of sphincterotomy and drainage. The use of intraductal direct cholangioscopy to visualize PBM has been reported by Mangas et al¹¹ without therapeutic purpose. The application of intraductal cholangioscopy for cancer surveillance of PBMs has shown encouraging results. Using the new digital system, Navaneethan et al¹² reported a sensitivity of 90.0% and a specificity of 95.8% for the diagnosis of malignancy based on visual impression. The sensitivity and specificity of cholangioscopy-targeted biopsies was 85% and 100%, respectively. The lower sensitivity of cholangioscopy-targeted biopsies compared with direct visualization of the lesion was probably related to the small

amount of tissue obtained with the miniature forceps used.^{12,13}

METHODS

A 16-year-old boy with recurrent pancreatitis was referred to our tertiary care center. He had undergone distal bile duct resection with Roux-en-Y hepaticojejunostomy for PBM, associated with Todani's IC choledochal cyst, 12 years earlier. Owing to recurrence of pancreatitis after surgery, in 2018 MRCP was used to confirm a long residual intrapancreatic choledochal stump (about 4 cm). The family initially refused a reintervention. Therefore, conservative management with large sphincterotomy and serial ERCP (approximately every 6-8 months) with protein plug removal and biliary stent placement was established. On the last ERCP (February 2021), a stable, eccentric minus image was noted, suggestive of a polyp. Direct cholangioscopy during ERCP with biopsies and recurrent stent placement was scheduled.

RESULTS

A standard duodenoscope was used ([Video 1](#), available online at www.giejournal.org). After 2 months, the biliary stent previously placed was already corroded by the pancreatic reflux. Virtual chromoendoscopy was used to assess the hypertrophic major papilla. A guidewire was left in place by using a 3-lumen sphincterotome, and cholangiography confirmed the suspected polyp. The sphincterotome was also used to confirm the patency of the papilla for cholangioscopy.

The cholangioscope (Spyglass DS II, Boston Scientific, Marlborough, Mass, USA) was introduced in the biliary stump. A water pump was used to facilitate distention of the lumen. The common channel and the pancreatic duct, with some protein plugs in a secondary duct, were visualized and washed. The surgical stump was reached and confirmed by the presence of nonabsorbable sutures ([Fig. 1](#)). The biliary polyp, 15 mm above the PBM and 1 cm under the stump, was evaluated, and a villous pattern was noted. Cholangioscope-guided biopsy with dedicated

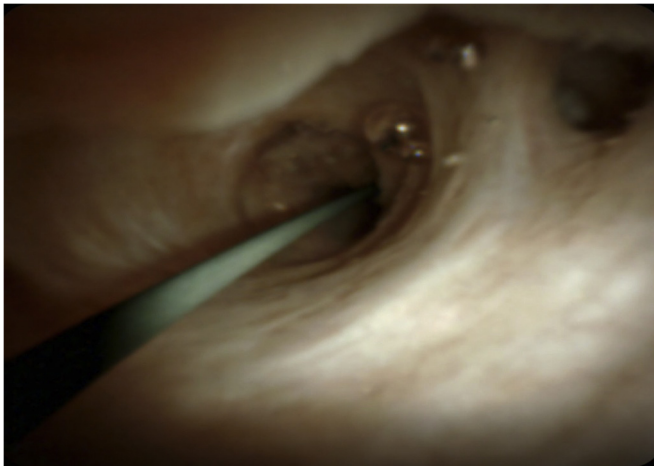


Figure 1. Cholangioscopy showing maljunction and the polyp in the biliary stump.

forceps and x-ray-guided biopsy with standard forceps were performed, and samples were stored separately. A 10F, 5-cm plastic stent was placed into the biliary stump to prevent stagnation and to facilitate tactile feeling of the intrapancreatic biliary stump in case of subsequent surgery.

The clinical course was uneventful. Histology revealed a monostratified cubic epithelium with mild inflammatory infiltration and the absence of atypia (hyperplastic polyp). Five months after stent placement, the patient was well with no episodes of pancreatitis. In consideration of the absence of neoplasm, we decided together with the family to postpone surgical reintervention and to continue endoscopic surveillance with x-ray-guided biopsies and stent replacement.

CONCLUSIONS

The management of isolated PBMs should be personalized and discussed in a multidisciplinary team. ERCP and direct cholangioscopy may play a role in surveillance, improving conservative management and ensuring direct access to bile duct lesions and better histologic assessment of potentially neoplastic lesions.

DISCLOSURE

All authors disclosed no financial relationships.

Abbreviation: PBM, pancreatobiliary maljunction.

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Digestive Endoscopy and Surgery Unit, Bambino Gesù' Children's Hospital, IRCCS, Rome, Italy (1), Sapienza University of Rome, Rome, Italy (2).

If you would like to chat with an author of this article, you may contact Dr Balassone at valerio.balassone@gmail.com.

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