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CASE IMAGE

Endoscopy and Procedures

Direct cholangioscopy supporting diagnosis and conservative surgery of a biliary rhabdomyosarcoma in a child

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Biliary rhabdomyosarcoma is the most common cause of malignant obstructive jaundice in pediatric patients.[1] After obtaining a prompt diagnosis, confirming the response to neoadjuvant chemotherapy can orientate liver-sparing surgery with favorable outcomes. Direct cholangioscopy is a well-established diagnostic and therapeutic technology in adults with an evolving role in children. We report a 3-year-old girl (15 kg) referred for unexplained jaundice despite an extensive workup. Magnetic resonance cholangiopancreatography demonstrated a right biliary dilatation and an aberrant anatomy. An endoscopic retrograde cholangiopancreatography (ERCP) was performed with cholangiogram, biliary sphincterotomy and a nasal-biliary tube placement for cytology that was not contributive. A computed tomography scan demonstrated a 4 cm × 2 cm mass in the right biliary duct, gallbladder, and common duct. Direct cholangioscopy confirmed intraluminal obliterative polypoid mass arising from the common duct without the involvement of the major papilla (Figure 1). Cholangioscopy-guided and X-ray-guided biopsies were taken and a plastic stent was left in place. Histology confirmed a well-differentiated embryonal biliary rhabdomyosarcoma positive to Desmin and Myogenin, with a high proliferation index (Ki67 40%). A tailored neoadjuvant chemotherapy according to European Pediatric Soft Tissue Sarcoma Study Group protocols was carried out for 3 months. Another ERCP with direct cholangioscopy was employed to confirm the tumor's response with a relevant mass reduction and to obtain negative biopsies from the left main duct and from the confluence (Figure 1). The patient received a right hepatectomy with "Roux-en-Y" hepaticojejunostomy. This report suggests a possible role for direct cholangioscopy even in small children for

[Correction added on 12 November 2024, after the first online publication: the affiliation 2 of Dr. Mario Brusco has been revised.]

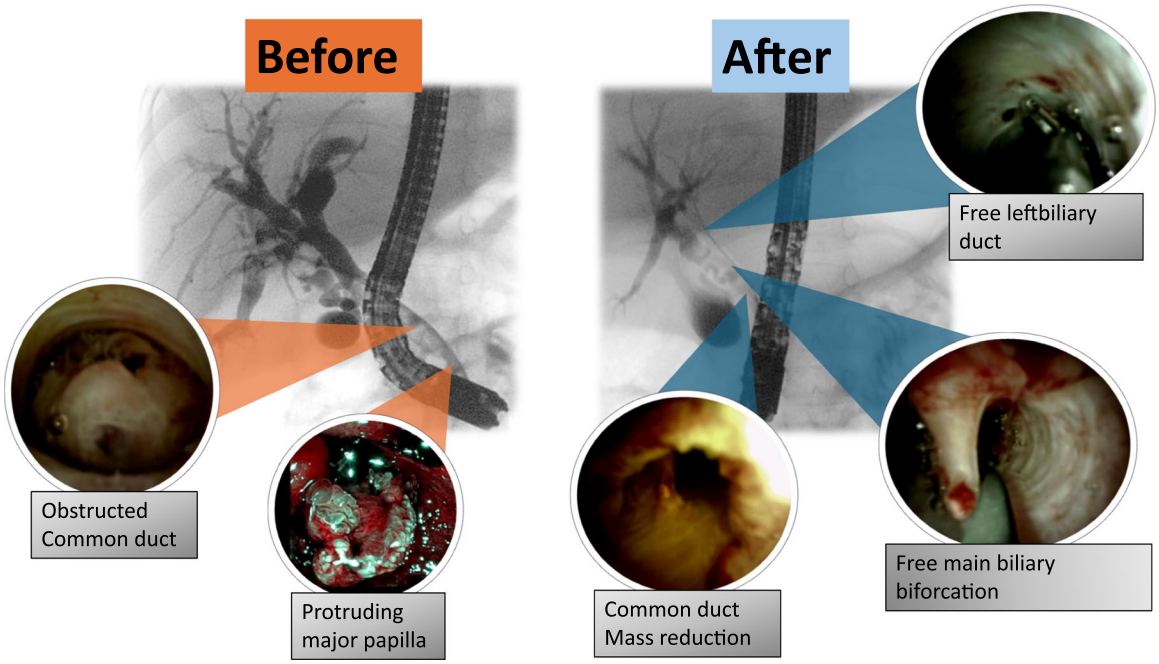

FIGURE 1 Cholangioscopy-aided diagnosis and evaluation of response after neoadjuvant chemotherapy to support the preservation of the left liver lobe.

difficult diagnosis or X-ray sparing endoluminal therapy.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

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