



Images in Surgery

Intrahepatic cystic mass and bile duct malformation

Arianna Pontrelli^{a,*}, Piercarmine Panzera^a, Francesco Paolo Prete^a, Enrico Fischetti^a,
Carlotta Testini^b, Mario Testini^a

^a Department of Precision and Regenerative Medicine and Jonian Area (DiMePRE-J), Academic General Surgery Unit "V. Bonomo", University of Bari, Italy

^b Department of Imaging Diagnostic, Radiodiagnostic Unit, University of Foggia, Italy

ARTICLE INFO

Keywords:

Biliary tract disease
Choledochal cyst
Hepatic hydatid cyst
Vascular abnormalities

A 62-year-old man was admitted to our department with persistent jaundice and diffuse abdominal pain lasting about a week. Laboratory tests presented hyperbilirubinemia (total bilirubin 8.03 mg/dL, direct bilirubin 5.59 mg/dL), increased gamma-glutamyl transpeptidase (296 U/L) and white blood cells ($18.68 \times 10^3/\mu\text{L}$), and negative tumor markers. Clinical history included diabetes mellitus, COPD and an untreated hepatic hydatid cyst diagnosed 30 years earlier. The patient had recently undergone ERCPs for recurrent cholangitis caused by microlithiasis. A previously performed emergency abdominal CT scan confirmed the hepatic cyst at the V-VII segment aside a gallbladder adenomyoma, choledocholithiasis and intrahepatic bile duct ectasia. Moreover, an appendicular formation suspicious for mucocele was diagnosed. The subsequent MRI described an ovoid mass-like structure of heterogenous signal intensity, compatible with echinococcal cyst, compressing the biliary confluence and causing dilatation of intrahepatic biliary tree (Fig. 1); the main bile duct was also dilated (13 mm) with multiple minute stones. Triphasic contrast CT scan documented dishomogenous lesion, 55 mm in diameter, sitting across the V-VII-VIII hepatic segments (Fig. 2); vascular reconstructions showed few anatomical variants: an aberrant right hepatic artery for the VI-VII segments branching from the superior mesenteric artery and an accessory left hepatic artery originating from the left gastric artery; furthermore, a right posterior venous branch for the VI-VII segments arose from the portal vein followed by intraparenchymal branching for the V-VIII segments, an anatomical variant known as type D/IV according to the classifications proposed by Nakamura [1] and Cheng. [2]. After a multidisciplinary meeting focused on Echinococcosis management, the

patient was started on oral *Albendazole* and then surgery was planned. At laparotomy, intraoperative ultrasound showed a 7 cm segment V hyperechoic mass appearing contiguous with a second 3 cm mass of the VI-VII segment, which displaced the vascular structures of the hepatic hilum. In consideration of the vascular variations previously diagnosed, a right posterior hepatectomy was performed to carefully approach the mass preserving as much hepatic parenchyma as possible. The procedure also included cholecystectomy, bile duct clearance and ileocecal resection for mucocele. Despite the patient's clinical history of hydatid cyst and the radiological characteristics of the cystic formation, intraoperative macroscopic aspect of the mass did not suggest an Echinococcal cyst but a cloacal dilatation of the bile confluence [3], filled with brown friable material and mucosal fragments. Postoperative pathology diagnosed a biliogenic suppurative hepatitis on end-stage liver cirrhosis with a focus of severe dysplasia of one of the bile ducts and an intracystic 7 mm poorly differentiated cholangiocarcinoma. Furthermore, the appendix was the site of a low-grade mucinous neoplasm. According to the *Todani* classification, which bile duct malformation should be considered?

- Type I
- Type III
- Type IVb
- Type V

* Corresponding author at: Department of Precision and Regenerative Medicine and Jonian Area (DiMePRE-J), University of Bari, Bari, Italy.

E-mail address: a.pontrelli9@studenti.uniba.it (A. Pontrelli).

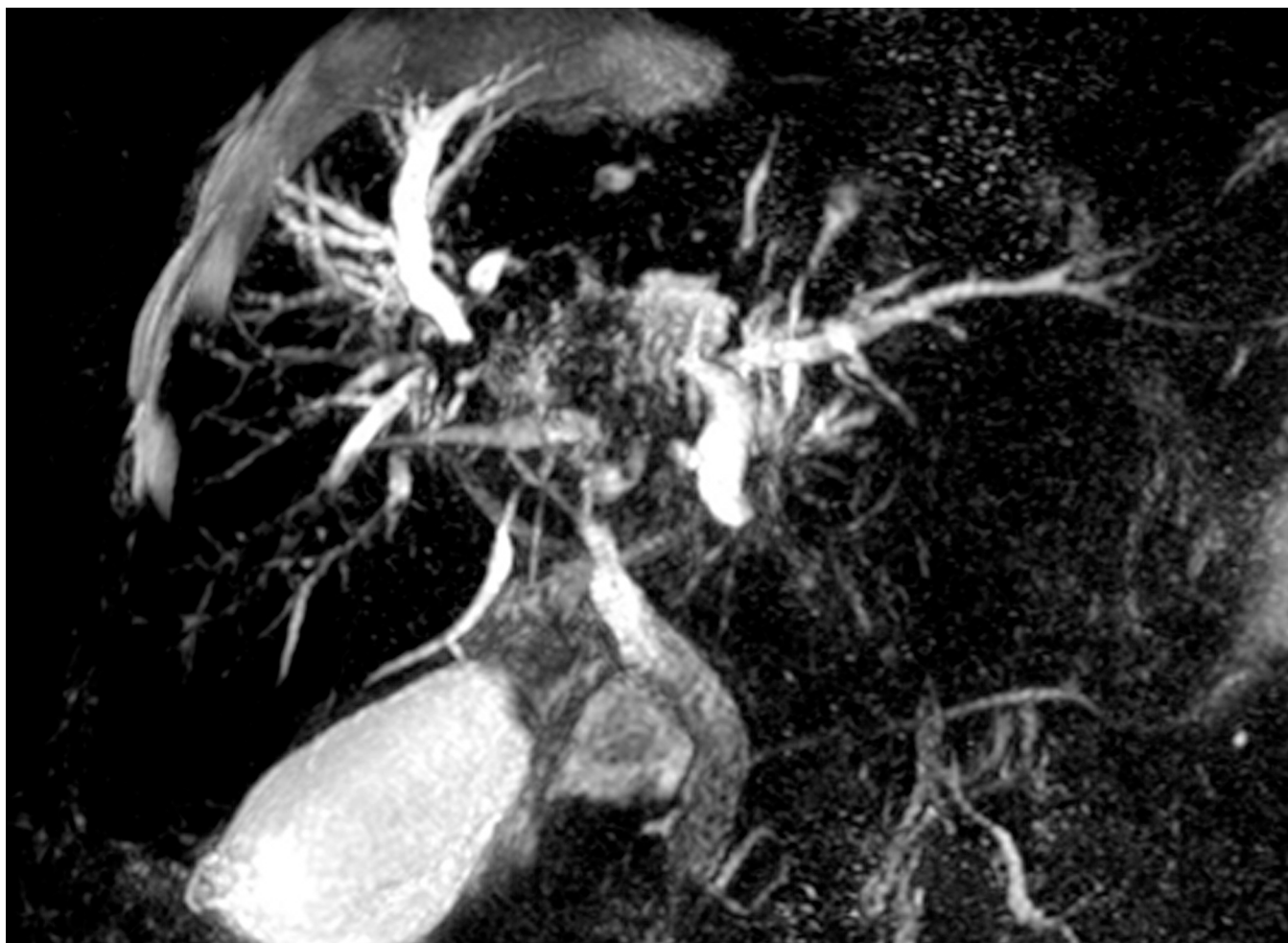


Fig. 1. Cholangiogram reconstruction of the biliary tree.



Fig. 2. Triphasic contrast CT scan showing a 55 mm dishomogenous mass across the V-VII-VIII hepatic segments.

Funding sources

This article did not receive any specific grant from funding agencies in public, commercial, or non-profit sectors.

Ethics approval

Not applicable.

CRediT authorship contribution statement

Arianna Pontrelli: Writing – review & editing, Writing – original draft, Conceptualization. **Piercarmine Panzera:** Visualization, Conceptualization. **Francesco Paolo Prete:** Writing – review & editing, Visualization. **Enrico Fischetti:** Visualization. **Carlotta Testini:**

Visualization. **Mario Testini:** Supervision.

Declaration of competing interest

All authors declare that they have no conflict of interest.

References

- [1] Nakamura T, Tanaka K, Kiuchi T, Kasahara M, Oike F, Ueda M, et al. Anatomical variations and surgical strategies in right lobe living donor liver transplantation: lessons from 120 cases. *Transplantation* 2002;73(12):1896–903.
- [2] Cheng YF, Huang TL, Lee TY, Chen TY, Chen CL. Variation of the intrahepatic portal vein: angiographic demonstration and application in living-related hepatic transplantation. *Transplant Proc* 1996;28(3):1667–8.
- [3] Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cysts: classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. *Am J Surg* 1977;134(2):263–9.