CASE REPORT

Cystic duct carcinoma mimicking a middle bile duct tumour

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SUMMARY

Cystic duct carcinoma was defined by Farrar as a tumour restricted to the cystic duct, making it a rare disease. The authors describe a case of a cystic duct carcinoma that fulfils Farrar's strict diagnostic criteria and that became clinically relevant by compressing the common hepatic duct, thus causing cholestasis. A cholecystectomy was performed with en bloc resection of the cystic and extrahepatic bile duct with a regional lymphadenectomy.

BACKGROUND

Cystic duct carcinoma was defined by Farrar in 1951 as a tumour restricted to the cystic duct,¹ making it a rare disease with an incidence of 0.9% of all malignant biliary tumours.² We present a case that fulfils the diagnostic criteria defined by Farrar: an early form of cystic duct carcinoma that compresses the common hepatic duct causing cholestasis. The final diagnosis was only achieved during surgery, given that the preoperative suspicion based on imaging was of a middle bile duct tumour.

CASE PRESENTATION

A 61-year-old woman with no remarkable medical history presented with a 3-day history of choloria, without abdominal pain or fever. Blood tests revealed total bilirubin 2.58 mg/dL, direct bilirubin 1.41, glutamate oxalacetate transaminase 223 U/L and glutamate pyruvate transaminase 595 U/L with negative inflammatory parameters. Abdominal ultrasonography showed dilation of the common bile duct and a distended gallbladder. MR cholangiopancreatography revealed a lesion of 7 mm in the common bile duct, 2.5 cm away from the hepatic confluence, with no invasion of the adjacent structures (figures 1 and 2). Owing to increased cholestasis, percutaneous biliary drainage was performed. Based on these findings, we established the presumptive diagnosis of a middle bile duct tumour. Laparotomy revealed gallbladder distension and a pericentimeter lesion in the cystic duct, near the junction with the common hepatic duct (figure 3). A cholecystectomy was performed with en bloc resection of the cystic, extrahepatic bile duct and lymph node dissection of the hepatoduodenal ligament. The biliary tract was reconstructed by a Roux-en-Y hepaticojejunostomy. Histological examination revealed a moderately differentiated adenocarcinoma of the cystic duct, with infiltration through the cystic wall, but with no invasion of the serosa, and with clear margins, staged pT2N0M0 (figure 4).

Figure 1 MR cholangiopancreatography revealing a lesion of 7 mm in the common bile duct, 2.5 cm away from the hepatic confluence (arrow).

OUTCOME AND FOLLOW-UP

The postoperative course was uneventful and the patient is doing well without any complication or recurrence 1 year from operation.

DISCUSSION

Cystic duct carcinoma was defined by Farrar in 1951 with the following diagnostic criteria: (1) the growth must be restricted to the cystic duct; (2) there must be no neoplastic process in the

Figure 2 Three-dimensional MR cholangiopancreatography sequence showing complete obstruction of the common bile duct.

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Figure 3 Resected specimen revealing a pericentimeter lesion in the cystic duct protruding to the common hepatic duct (arrows).

gallbladder, hepatic or common bile duct and (3) histological examination of the growth must confirm the presence of carcinoma cells in the mass.¹ The small number of cases reported is explained by the fact that these strict criteria can only be applied to early cystic duct carcinoma, as the growth of the tumour with invasion of adjacent structures surpasses this definition. Different classifications have been proposed using an alternative definition, based on the location of the tumour centre, making it more frequent with an incidence in 5.6–6.6% of all malignant biliary tumours.^{2–3} In contrast to gallbladder carcinomas, it is more frequent in males,² and gallstones are less associated with it, being found in only 25.7% of all patients.⁴ Patients can present abdominal pain (68.9%), caused usually by gallbladder hydrops or cholecystitis, and obstructive jaundice (34.3%) due to proximity of the cystic duct to the common hepatic duct.⁴ The symptoms are common with other hepatobiliary diseases, so CT and MR cholangiopancreatography are crucial to diagnose and determine resectability. The standard surgical procedure for an early cystic duct carcinoma is a com-

bined excision of the gallbladder, and cystic and extrahepatic bile duct with lymph node dissection of the hepatoduodenal ligament.⁵ The more advanced forms will need a combined resection with hepatectomy or pancreatoduodenectomy because of perineural and lymphatic infiltration.^{2–3} Cystic duct carcinoma has a better prognosis than that of other biliary ducts and gallbladder carcinoma, explained by a higher resectability, since it is expected to cause symptoms at an early stage because of the obstruction of the cystic duct.⁴ Reported 5-year survival is 22–40% with a median survival of 1.5–2.4 years.^{2–6}

Learning points

- Cystic duct carcinoma, defined by Farrar, is a tumour restricted to the cystic duct, making it a rare disease.
- It shares the same clinical presentation with other hepatobiliary diseases, increasing the importance of imaging.
- In its early form it has a better prognosis, and the standard surgical procedure is a combined excision of the gallbladder, and cystic and extrahepatic bile duct with lymph node dissection of the hepatoduodenal ligament.

Figure 4 Adenocarcinoma infiltrating through the muscular layer into the subserosal tissue (H&E ×20).

Competing interests None.

Patient consent Obtained.

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