Carcinoid Tumor of the Cystic Duct: Case Report, Literature Review and Comparison of Surgical Approaches

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Key Words
Carcinoid · Cystic duct · Laparoscopic resection

Abstract
We present the case of a 48-year-old female with a cystic duct carcinoid that was found incidentally upon laparoscopic cholecystectomy. The patient subsequently underwent excision of the cystic duct remnant. This is the tenth reported case of a cystic duct carcinoid and the second reported laparoscopic excision.

Introduction
Carcinoid tumors of the gastrointestinal (GI) tract are rare, frequently indolent and have a high survival rate without metastasis [1]. They are variable and present with nonspecific symptoms, thus making a diagnosis difficult [1]. An extremely rare site of GI carcinoid tumor is the extrahepatic biliary tree, harboring 0.1–0.2% of all GI tumors [2]. In the extrahepatic biliary tree, cystic duct carcinoids represent 11% [3] of all reported cases.

Case Report
A 48-year-old female presented with right upper quadrant and epigastric pain, also complaining of nausea, vomiting and diarrhea. She was first examined in the emergency room, where a sonogram identified a normal gallbladder with no evidence of gallstones, no thickening of the gallbladder wall and no pericholecystic fluid. A Disida scan identified a
delayed visualization of the gallbladder and small bowel, which is consistent with biliary dyskinesia. A liver panel was ordered and the total bilirubin was 2.2 mg/dl (normal range: 0.3–1.5), ALT (SGPT) was 475 IU/l (normal range: 5–60), AST (SGOT) was 630 IU/l (normal range: 15–43) and the alkaline phosphatase was 158 IU/l (normal range: 34–121).

The patient’s case was complicated as she was being treated for complex Lyme’s disease with co-infections of babesiosis and Bartonella. She was taking Factive (fluoroquinolone), azithromycin, minocycline and IV Rocephin (a ceftriaxone antibiotic). Gallbladder disease symptoms have been reported with the use of ceftriaxone [4].

A follow-up liver panel performed 8 days after emergency room consultation was within normal limits. Follow-up ultrasound performed 2 weeks after emergency room consultation identified what appeared to be 1–2 stones in the neck of the gallbladder.

A laparoscopic cholecystectomy was recommended and performed uneventfully, and the patient did well postoperatively. The surgical pathology identified an incidental carcinoid tumor in the cystic duct measuring up to 2.5 mm without a negative margin. The tumor cells were positive for keratin, synaptophysin and chromogranin.

The identification of the carcinoid tumor led to further evaluation and imaging, which was done prior to the reoperation to remove the remaining tumor and obtain clear margins. An MRI of the abdomen with and without contrast identified a 13-mm remnant of the cystic duct. Imaging revealed no obvious residual carcinoid or metastatic disease. Surgery was performed with laparoscopic removal of the cystic duct remnant and the pathology report identified a cystic duct remnant with no evidence of residual carcinoid tumor. At follow-up by a medical oncologist, the patient had a normal serum serotonin level. At a subsequent follow-up 4 years after the laparoscopic excision, the patient had a normal physical examination and normal liver ultrasound with no evidence of metastasis.

**Discussion**

A literature review identified only 9 previously reported cases of cystic duct carcinoids (table 1). Of these, only 1 case was reported to be laparoscopically excised.

A carcinoid of the cystic duct arises from enterochromaffin cells, which are neuroendocrine cells of the GI tract [5]. These tumors have been found to be more prevalent in women than in men (8 out of 10 cases) [3, 6]. Patients’ ages range from 28 to 69 years [2, 3], and the most common preoperative complaint is right upper quadrant abdominal pain [3, 6]. All cystic duct carcinoids reported were found incidentally upon laparotomy or laparoscopy with pathologic analysis.

As there are so few cases reported of cystic duct carcinoids, the surgical standard of care has yet to be established. Laparoscopic or open surgery is performed to obtain clear margins. In the only other reported laparoscopic removal of the cystic duct carcinoid [3], a Roux-en-Y hepaticojejunostomy was performed in conjunction with the excision of a large mesenteric lymph node. In this case, the patient had locoregional disease.

We propose that if the cystic duct carcinoid is small and there is no common duct involvement, a complete laparoscopic resection of the cystic duct can be performed without injuring the common bile duct (fig. 1). This obviates the need for common bile duct excision and reconstruction of the biliary tree. Laparoscopic resection is possible if negative margins are achieved and no involvement of the regional lymph nodes can be confirmed. Metastasis to the regional lymph nodes and the liver are the most common sites reported [2, 6]. In cystic duct carcinoids, metastasis is infrequent, occurring in 2 of the 10 cases [2].
In conclusion, carcinoid of the cystic duct is quite rare and difficult to diagnose. Thus far, surgical resection is the primary modality of treatment [2]. Our case demonstrates the excellent outcome of a laparoscopic resection (fig. 2). In suitable patients, it can be performed to obtain clear margins, obviating the need for common bile duct resection or complex biliary tree reconstruction.

Disclosure Statement

The authors have no conflicts of interest to declare.

References


Table 1. Patient data of reported cystic duct carcinoids

<table>
<thead>
<tr>
<th>Case</th>
<th>Authors [ref.], year</th>
<th>Age</th>
<th>Sex</th>
<th>Preoperative complaint</th>
<th>Metastasis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Goodman et al. [7], 1984</td>
<td>28</td>
<td>F</td>
<td>Right upper quadrant pain</td>
<td>Lymph node</td>
</tr>
<tr>
<td>2</td>
<td>Nicolescu and Popescu [8], 1986</td>
<td>50</td>
<td>F</td>
<td>Right upper quadrant pain</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>Chittal and Ra [9], 1989</td>
<td>46</td>
<td>F</td>
<td>Right upper quadrant pain</td>
<td>None</td>
</tr>
<tr>
<td>4</td>
<td>Rugge et al. [10], 1992</td>
<td>64</td>
<td>M</td>
<td>Jaundice</td>
<td>None</td>
</tr>
<tr>
<td>5</td>
<td>Meyer et al. [11], 1997</td>
<td>56</td>
<td>F</td>
<td>Right upper quadrant pain</td>
<td>None</td>
</tr>
<tr>
<td>6</td>
<td>Shah et al. [12], 1998</td>
<td>52</td>
<td>F</td>
<td>Right upper quadrant pain</td>
<td>None</td>
</tr>
<tr>
<td>7</td>
<td>Hermina et al. [13], 1999</td>
<td>69</td>
<td>M</td>
<td>Right upper quadrant pain</td>
<td>Lymph node</td>
</tr>
<tr>
<td>8</td>
<td>Felekouras et al. [2], 2007</td>
<td>60</td>
<td>F</td>
<td>Jaundice</td>
<td>None</td>
</tr>
<tr>
<td>9</td>
<td>Lim et al. [3], 2012</td>
<td>58</td>
<td>F</td>
<td>Right upper quadrant pain</td>
<td>None</td>
</tr>
<tr>
<td>10</td>
<td>Present case, 2014</td>
<td>48</td>
<td>F</td>
<td>Right upper quadrant pain</td>
<td>None</td>
</tr>
</tbody>
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Fig. 1. Cystic duct remnant retracted.

Fig. 2. Cystic duct remnant (long arrow). Common bile duct retracted (short arrow).