Carcinoid of the Appendix During Laparoscopic Cholecystectomy: Unexpected Benefits

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ABSTRACT

Carcinoid tumors of the midgut arise from the distal duodenum, jejunum, ileum, appendix, ascending and right transverse colon. The appendix and terminal ileum are the most common location. The majority of carcinoid tumors originate from neuroendocrine cells along the gastrointestinal tract, but they are also found in the lung, ovary, and biliary tracts. We report the first case of elective laparoscopic cholecystectomy in which we found a suspicious lesion at the tip of the appendix and proceeded to perform a laparoscopic appendectomy. The lesion revealed a carcinoid tumor of the appendix.

Key Words: Carcinoid, Tumor, Appendix, Laparoscopy.

CASE REPORT

A 46-year-old female presented with vague gastrointestinal symptoms, chronic right upper quadrant abdominal pain and intolerance to fatty food and diarrhea for the last two years. She was given the diagnosis of irritable bowel syndrome and had been followed by her internist. She had undergone an abdominal hysterectomy with bilateral salpingo-oophorectomy for menometrorrhagia and fibroids 15 years ago but was otherwise healthy. During the work-up, an ultrasound (U/S) of the abdomen revealed several small stones in the fundus of the gallbladder and a common bile duct of normal diameter. The appendix was not visualized in the U/S. She was then scheduled to undergo an elective laparoscopic cholecystectomy.

Pneumoperitoneum was established with CO₂ gas insufflation under general anesthesia in the usual fashion. Laparoscopic visualization of the abdominal cavity revealed a small brownish mass at the tip of the appendix resembling a lymph node. The gallbladder was removed via the standard American laparoscopic technique (4 ports). The camera was then moved from the supraumbilical port to the subxyphoid one. Using the same instruments, ie, two Endo grasps plus a Multifire Endo GIA 30 stapler, the appendix was removed by stapling across the base of the appendix and across the mesoappendix. Pathologic examination demonstrated a carcinoid tumor in the tip of the appendix, which measured 0.3 x 0.2 cm (Figure 1, 2). The margins of the specimen were clear, no lymph nodes were seen, and there was no mesoappendix invasion. The patient was discharged 23 hours after the operation. She remains asymptomatic at six months follow-up visit.

DISCUSSION

Merling, in 1838, described the gross pathology of carcinoid tumor of the appendix, and in 1907 Oberndorfer coined the term “Karzinoide.”¹² Carcinoid tumors are the most common gut endocrine tumors.³ They arise from the foregut, midgut, and the hindgut. Carcinoid tumors of the midgut arise from the distal duodenum, jejunum, ileum, appendix, and ascending and right transverse colon, with the appendix and terminal ileum being...
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Godwin reviewed 2,837 cases of carcinoid tumor and found that the majority originate from the gastrointestinal tract (85%), but they were also found in the lung, ovary, and biliary tracts. Most were in the appendix, rectum, and ileum. Age-adjusted incidence rates were higher for black males.

Carcinoid tumors showed several differences from other kinds of tumor, including a low age for appendiceal and lung cases and low male/female and black/white ratios in the lung. Godwin's percentages of concurrent neoplasm and multiple carcinoids were low compared to other series.

Survival varies with anatomical location and histological characteristics. Five-year relative survival rates ranged from 99% (appendix) to 33% (sigmoid colon). However, survival for colon cases was not as low as expected on the basis of the high rate of metastases. Histologically, there are five generally accepted carcinoid growth patterns, ie, insular (type A), trabecular (type B), glandular (type C), undifferentiated (type D), and mixed type. The mixed tumors with acinar and glandular pattern have the best median survival time (4.4 years), whereas the undifferentiated has the poorest median survival (0.5 year). In decreasing order of median survival time in years, the growth patterns ranked as follows: mixed insular plus glandular, 4.4; insular, 2.9; trabecular, 2.5; mixed insular plus trabecular, 2.3; three pooled low incidence rate mixed growth patterns, 1.4; glandular, 0.9; and undifferentiated, 0.5.

The site of origin and rate of metastases varies, with appendix being the most common site of carcinoid. The frequency of metastasis from carcinoid of the appendix varies between 1.4% and 8.8%. Carcinoids of the appendix and rectum smaller than 2 cm have a 5-year survival rate of approximately 100%, declining to 40% when the tumor diameter is larger than 2 cm. However, metastases have been reported from carcinoid of the appendix less than 2 cm in size. While the risk of metastasis of these tumors has been correlated with their size, invasion of the mesoappendix is predictive of an increased risk of metastasis for carcinoid tumors of the appendix less than 2 cm in size. Tumors with liver metastases have a 5-year survival rate ranging from 21% to 42%

The clinical presentation of carcinoid tumors depends on their location with a wide clinical spectrum varying from asymptomatic to the malignant carcinoid syndrome. Carcinoids of the appendix and small intestine are slow-
growing and silent. They are, in most instances, found incidentally at operation, as in our case. A third of patients with carcinoid present with years of intermittent abdominal pain often ascribed to other gastrointestinal or biliary disease, or to the irritable bowel syndrome. Malignant carcinoid tumors may present with symptoms of mechanical bowel obstruction due to fibrosis, adhesions and kinking of the intestine. Other constitutional symptoms include weight loss, diarrhea, upper gastrointestinal bleeding, intussusception, and abdominal mass. In patients with carcinoid of the foregut (atypical carcinoid), the urine contains only slightly elevated levels of 5-hydroxyindoleacetic acid (5-HIAA), but large quantities of 5-hydroxytryptophan (5-HTP) and 5-hydroxytryptamine (5-HT), because these tumors are deficient in dopa-decarboxylase, which is responsible for the conversion of 5-HTP to 5-HT. The excess 5-HTP in the plasma is directly excreted into the urine. In carcinoid of the midgut (typical carcinoid) most of the 5-HTP is rapidly converted to 5-HT, which is taken up by platelets. The excess of 5-HT is converted by monoamine oxidase (MAO) and aldehyde dehydrogenase (DA) to 5-HIAA, which then is excreted in high concentrations in the urine. It is interesting to note that serotonin-rich foods, such as bananas, plantains, pineapples, kiwi fruits, walnuts, hickory nuts, pecans, avocados, and acetaminophen may artificially elevate 5-HIAA.8

Numerous techniques can identify the primary site of the tumor. A chest radiograph or computed tomography (CT) may show a bronchial or mediastinal tumor17,18 Double-contrast gastrointestinal studies still best define the primary neoplasm. Appendiceal tumors frequently escape radiological detection until large enough to be discovered by computed tomography (CT). The hypervascular nature of carcinoid tumors and their metastases allows superior mesenteric arteriography of the small bowel and cecum to be used when the scanning procedures are not revealing. The “spokewheel” configuration of the desmoplastic mesenteric masses and lymph node metastases are best seen by CT, whereas hepatic metastases can be demonstrated by CT, CT-angioportography (CTAP), ultrasonography (US), magnetic resonance imaging (MRI), and octreotide scintigraphy. Percutaneous needle biopsy with radiological guidance may confirm the diagnosis of carcinoid tumors and their metastases. Hepatic arteriography is frequently performed in preparation for hepatic embolization or chemoembolization.19,20 Early resection offers the patient the best chance of cure. Carcinoid should be resected regardless of the presence of metastases.

Appendiceal carcinoid tumors are rarely malignant, and lesions smaller than 1.5 cm can be safely resected by appendectomy only. If the tumor is at the base of the appendix, a cecectomy may prove necessary. Tumors larger than 1.5 cm require an ileocectomy.21,22 Laparoscopic resection of carcinoid tumors from the stomach,23 gallbladder,24 proximal25 and distal26 rectum have been described. Laparoscopic appendectomy has been compared to open procedures, and the results showed open procedures taking less time, but there were more wound infections than in the laparoscopic procedure. Patients with acute appendicitis recuperated more quickly from the laparoscopic procedure, as evidenced by the time until eating regular diet, period of hospitalization, incidence of nausea and pain medications on postoperative day one.27-31

CONCLUSION

We report a case of incidental laparoscopic resection of a carcinoid tumor at the tip of the appendix during an elective laparoscopic cholecystectomy. Even though the risk of metastasis from carcinoid correlates with the size, ie, greater than 2 cm, there are several cases of metastatic disease from carcinoid tumors smaller than 1 cm in diameter. Based on review of the literature, we suggest that any suspicious lesion of the appendix found during laparoscopic exploration of the abdominal cavity should be removed. We believe that the magnification provided by the laparoscope, ie, up to 18 times, and the ability to inspect the abdominal cavity allowed us to identify the lesion and to perform laparoscopic appendectomy while adding minimal morbidity to the procedure.

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