Carcinoid tumor of bile duct

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We report a 38-year-old lady with carcinoid tumor of the extrahepatic biliary tract who presented with recurrent obstructive jaundice and previous surgery for suspected cholelithiasis. MRCP revealed a large bile duct tumor extending from the confluence up to the superior aspect of the pancreas; this was completely excised, with bilio-enteric anastomosis. These tumors are characteristically slow growing and, therefore, are amenable to aggressive surgical excision, which offers the best chance of cure. [Indian J Gastroenterol 2005;24:262-263]

Carcinoid tumors of the biliary tract are rare and account for 0.2%-2% of all GI carcinoids. These are usually slow growing neoplasms with low malignant potential and hence are amenable to aggressive surgical management. Preoperative diagnosis is difficult because they mimic the signs and symptoms of cholelithiasis and/or cholangiocarcinoma.

A 38-year-old lady presented to a surgeon three years ago with history of obstructive jaundice, pain, fever, and serum bilirubin of 8 mg/dL. She underwent endoscopic retrograde cholangiography (ERC), which revealed hemobilia; biliary stenting was done. Following ERC, serum bilirubin dropped to 1.4 mg/dL and the patient was asymptomatic. She presented again with obstructive jaundice a year later and underwent cholecystectomy with removal of biliary stent. Intraoperatively, a suspicious mass was noticed in the common bile duct (CBD); this was biopsied and a T-tube was inserted. Histology was reported as neuroendocrine tumor, which was confirmed by immunohistochemistry (cytokeratin and leukocyte common antigen [LCA] positive).

The patient followed up intermittently thereafter. A year later, CT abdomen revealed a lesion at the confluence encasing the plastic stent, with intrahepatic biliary radicle (IHBR) dilatation. She underwent palliative stenting. One year later she presented with obstructive jaundice since 2 months. ERC was done and previous stent was removed; due to bleeding, a new stent could not be placed.

She was then referred to our department. CT scan revealed a mass involving the extrahepatic biliary tract from confluence to superior border of pancreas. Fat planes between the mass and duodenal wall and head of pancreas were obliterated superomedially, with mildly prominent IHBR. Magnetic resonance cholangio-pancreatography (MRCP) showed an enhancing mass lesion, 7 cm x 4 cm x 3 cm, extending from the confluence of the hepatic ducts to the distal CBD (Fig). The mass involved the entire CBD and extended into the left hepatic system with IHBR dilatation. The right duct measured 12 mm and left 14 mm.

Preoperative percutaneous transhepatic biliary drainage (PTBD) was done to facilitate identification of the confluence and biliary ducts. Intraoperative findings revealed a large, firm, nodular tumor of the CBD measuring 5 cm x 4 cm x 3 cm, extending from the suprapancreatic part of the CBD up to the confluence. Total excision of the tumor was carried out with a margin of 0.5 cm above and up to the upper border of pancreas. Continuity was established with a Roux-en-Y hepatico-jejunostomy. Postoperative period was uneventful. PTBD was clamped and removed after 12 days.

Histology confirmed the tumor to be a well-differentiated neuroendocrine carcinoma of the CBD. One lymph node was free of tumor. Both the resection margins were free. The tumor showed a combination of insular, acinar and trabecular pattern. Mitotic activity was seen (6-10 mitosis/HPF). The tumor was seen invading through the wall of CBD.

The patient recovered uneventfully and is asymptomatic on follow up at 2 months.

Carcinoid tumor of the extrahepatic bile duct is a rare form of biliary obstruction, with fewer than 34 cases reported in the English literature. The most common anatomic sites were the CBD (58%), perihilar region (28%), cystic duct (11%), and common hepatic duct (3%). Unlike cholangiocarcinoma, biliary carcinoids occur more commonly in younger patients and in women. Aggressive local invasion by the primary tumor is rare, and metastases occur in less than one-third of patients. Surgical resection is recommended. The final diagnosis is usually confirmed by immuno-histochemistry studies.

Data on adjuvant therapy remain investigational;

Fig: MRCP showing upper extent of large tumor with dilated right and left ducts

Case Snippets
however, available information suggests that patients with biliary carcinoid have an overall favorable prognosis after aggressive surgical management. The mean disease-free follow up after resection in one study was 32 months (range, 3 months-20 years). Hence surgery with curative intention should be considered wherever possible.

References

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Thyrotoxicosis co-existing with ulcerative colitis

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A 32-year-old lady was admitted with complaints of recurrent episodes of vomiting and loose stools associated with tenesmus and blood and mucus. She also had a neck swelling since 11 years of age, and had features of thyrotoxicosis for five to six months. She responded to balsalazide only after her thyroid status was controlled with carbimazole. [Indian J Gastroenterol 2005;24:263-264]

Autoimmune thyroid diseases are well known to complicate ulcerative colitis, but the association of thyrotoxicosis with ulcerative colitis is not common. Control of active colitis is possible only after treatment of hyperthyroidism.

A 32-year-old lady presented with complaints of recurrent vomiting and bloody diarrhea with tenesmus. There was no significant past medical illness. Examination showed an anxious patient with tachycardia and exophthalmos. There was a multinodular goiter with no bruit. Abdominal examination was normal.

Investigations showed leukocytosis with elevated ESR. Colonoscopy showed active ulcerative colitis involving the left side of the colon; this was confirmed on histology. Thyroid profile showed undetectable serum TSH with elevated free T3 and T4 levels. Technetium-99m thyroid scintigraphy showed increased uptake in both the lobes suggestive of a large multinodular goiter. Anti-thyroid antibodies were negative. She was treated with balsalazide and topical steroids, with no improvement for the initial 7 days. She was then started on carbimazole and low-dose propranolol. A week later she showed improvement in her thyrotoxic and bowel symptoms and was in complete remission of ulcerative colitis when reviewed one month later. She is in remission on maintenance dose of balsalazide and carbimazole for the last 6 months.

Though autoimmune thyroid disorders are well known to complicate chronic ulcerative colitis, the coexistence of hyperthyroidism and ulcerative colitis is very rare. The incidence of thyrotoxicosis in patients with ulcerative colitis is 0.8% to 3.7% and the prevalence of ulcerative colitis in patients with hyperthyroidism is 1.3%. Some investigators have suggested only an incidental association of these two diseases, while others reported a relationship between the two. Psychological stress may exacerbate either disease.

Hyperthyroidism intensifies the systemic manifestations of ulcerative colitis and renders its management difficult. The rapid metabolism of the drugs for treating ulcerative colitis or their rapid transit through the gut may prevent them from attaining effective concentrations. Hence before the colitis can be effectively controlled, treatment of the thyrotoxicosis is essential.

Silent hyperthyroidism in ulcerative colitis has also been reported. Also some of the features of active colitis may obscure an early diagnosis of coexisting thyrotoxicosis. Hence laboratory tests for thyrotoxicosis are indicated in patients with refractory ulcerative colitis with or without thyroid enlargement.

Either one of the diseases in active stage can induce the other. Since our patient had a toxic multinodular goiter, the chance of other nodules turning autonomous in the future is high. So the treatment of choice is total thyroidectomy with thyroid hormone replacement.