Carcinoid of ampulla of Vater

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Abstract
Carcinoid tumors of ampulla are rare clinical entities. They form 0.35% of all the gastrointestinal carcinoids. So far, only 109 cases have been reported in the literature, mostly as individual case reports. Since the metastatic potential and the tumor size have no correlation, unlike in duodenal carcinoids, pancreatoduodenectomy is considered the treatment of choice. Here we present a case of carcinoid of ampulla presenting to our department.

Key words: Ampulla of Vater, carcinoid, Whipple’s operation

Introduction
Carcinoid tumors of ampulla are rare and form less than 0.35% of gastrointestinal carcinoids.[1] These arise from enterochromaffin cells, also known as argentaffin cells or kultchitski cells, a part of amine precursor uptake and decarboxylation (APUD) system. These are distributed in GI tract, pancreas and bronchi. In the GI tract, they are most commonly localized to small intestine, appendix and rectum. Carcinoid tumors of GI tract most commonly present with abdominal pain, bleeding, obstruction and symptoms associated with tumor-secreted hormones.

Case Report
A 24-year-old woman presented to our department with pain in abdomen, malena and mild jaundice. On examination, she had pallor. Her hemoglobin was 9 gm%. Serum bilirubin was 3 mg% (direct 1.9 mg%, indirect 1.1 mg%), alanine transaminase 61 IU/L, aspartate transaminase 145 IU/L and alk phosphatase 261 U/L.

Ultrasonography showed a hypoechoic mass in the ampulla projecting into second part of duodenum with dilated common bile duct and intrahepatic biliary radicles. A few enlarged para-aortic nodes were seen.

Upper GI endoscopy showed a growth involving the ampulla with normal duodenal walls. Biopsy was taken. It was suggestive of neuroendocrine tumor. CT scan revealed a mass involving the ampulla with enlarged gallbladder and dilated CBD and intrahepatic biliary radicles [Figure 1].

The patient underwent laparotomy; there was no metastasis in the liver. Pancreatoduodenectomy (Whipple’s operation) with lymph node dissection was performed. The reconstruction involved end-to-end pancreaticojejunostomy, hepaticojejunostomy and gastrojejunostomy. The post-operative period was uneventful, and the patient was discharged the 15th day.

Histopathological examination revealed a tumor in the ampullary region of 2.2 cm in size. The tumor invaded the muscularis layer. The tumor showed a...
predominantly insular pattern of growth with areas of focal necrosis [Figure 2]. Four out of 27 nodes showed metastatic tumor. Immunohistochemistry was positive for neuron specific enolase (NSE) and chromogranin and epithelial membrane antigen.

Discussion

The first case of carcinoid was described by Lubarsch in 1888 in the ileum. Oberndorfer coined the term carcinoid in 1907. Williams and Sandler classified carcinoids according to site of origin as foregut, midgut and hindgut carcinoids.[2] Carcinoids most commonly arise in the small intestine, bronchus, appendix and rectum.[3] Midgut carcinoids are argentaffin positive, have high serotonin content and secrete hormones to cause carcinoid syndrome when they metastasize. Carcinoid syndrome is due to the release of serotonin and tachykinins.[4]

Carcinoids can be classified histologically into five different types: insular, trabecular, glandular, undifferentiated and mixed. Midgut carcinoids most typically present with insular morphology. Foregut carcinoids show a mixed pattern, and hindgut carcinoids are frequently solid or trabecular.[5] Immunohistochemical analysis is done by identification of chromogranin, synaptophysin and NSE.[6,7]

Ampullary carcinoids form around 2% of all ampullary tumors and 0.35% of all gastrointestinal carcinoids. In the literature, 109 cases have been described, out of which around 60 cases are associated with neurofibromatosis type-1 (NF-1). This strong association with NF-1 was described by Kline et al.[8] Perrone attributed this to ectodermal-endodermal complex transformation, which is situated in the ampulla of Vater.[9] Patients present with neurofibromatosis with abdominal pain and jaundice should be thoroughly screened for ampullary tumor.[8]

The most common presentation is painless jaundice followed by pain, bleeding (3%) and pancreatitis. Carcinoid syndrome is a rare manifestation; only two cases are found in the literature so far.[10,11] Diagnosis is established by USG, CT scan and endoscopy with biopsy. These could be supplemented with MRCP. Selective venous sampling, positron emission tomography (PET) and various forms of radionuclide scanning [radiolabelled somatostatin receptor scintigraphy (SRS) and iodinated metaiodobenzylguanidine (MIBG) scanning] can be used to localize the tumor.[12]

It has been shown that size of tumor has no correlation with the metastatic potential unlike with duodenal carcinoids where tumor size, invasion of muscularis propria and mitotic activity correlated with metastatic potential. Duodenal tumors less than 2 cm can be locally resected. In contrast, for ampullary carcinoids, the size and mitotic activity have no prognostic significance.[13] Even tumors less than 1 cm can present with metastasis, and local resection has high propensity to leave behind node positive tissue.[11]

Several authors have recommended local resection for ampullary carcinoids, irrespective of the size. But, these recommendations were followed when Whipple's procedure carried a high mortality (around 25%). In the present era of expertise and high safety in Whipple's operation, and given the high propensity for even small tumors to present with nodal metastasis, pancreatoduodenectomy is considered the operation of choice.[11]

References


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