

Case Report

Rare Association of Ampullary Carcinoma

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Abstract: Ampullary carcinoma is the second most common cancer of the ampullary area after pancreatic carcinoma. Though ampullary carcinoma is known to metastasize intra-abdominally and to the liver, solid adrenal metastasis has not been reported so far, to the best of our literature search. Here we describe a patient with ampullary carcinoma, solid adrenal metastasis and incidental coexistent aortic aneurysm.

Keywords: Ampullary carcinoma, Adrenal Metastasis, Intrahepatic biliary radical (IHBR) dilatation, Aneurysm

INTRODUCTION:

Ampullary cancer is a rare gastrointestinal cancer which arises from the tip of the common bile duct and passes through the wall of the duodenum into ampullary papilla. A number of different abnormal conditions may affect the ampulla, including obstruction from bile duct stones, localized or generalized inflammatory processes, trauma, and neoplastic diseases [1]. According to the American Cancer Society (ACS), almost everyone who has ampullary cancer has jaundice, hence it is often found at an earlier stage than pancreatic cancer [2].

CASE REPORT:

A 60 year old gentleman came with complaints of abdominal pain, loss of weight and appetite for 6 months and jaundice for 4 months. He had complaints of itching all over the body on and off for one week. There was no history of vomiting / hematemesis. He gave history of tarry stool 2 months back, which lasted for about 10 days. On examination, patient's general nutrition was poor. Patient appeared anemic and icteric and had a palpable lump in the right hypochondrium. In lab investigations, patient's total bilirubin was 7.3 mg/dl.

Plain radiograph showed cardiomegaly with left ventricular configuration and unfolded prominent aorta (Fig: 1).



Fig 1: Plain X-ray chest PA shows cardiomegaly with LV configuration and unfolded prominent aorta.

Plain and CECT abdomen revealed gross IHBR dilatation. Common bile duct and main pancreatic duct were dilated throughout its course and measured ~14 mm at porta hepatis [Fig: 5(a) & (b)].

Small enhancing nodular lesion measuring ~ 11 x 10 mm noted in the region of ampulla – Ampullary growth [Fig: 2(a)]. Enhancing lesion noted in left adrenal gland – Metastasis [Fig: 4].

Associations:

Focal dilatation noted in the distal thoracic aorta for a segment of ~ 5 cm with a thrombus noted along the anterior aspect within the dilated segment causing about 30% stenosis – Fusiform aneurysm of thoracic aorta [Fig: 2(a)&(b)]. A well-defined thick

walled hypo dense lesion (HU 10 – 16) with thin septations and no significant enhancement was noted in segment VIII of liver - Complex cyst / hydatid cyst [Fig: 3(a)&(b)].

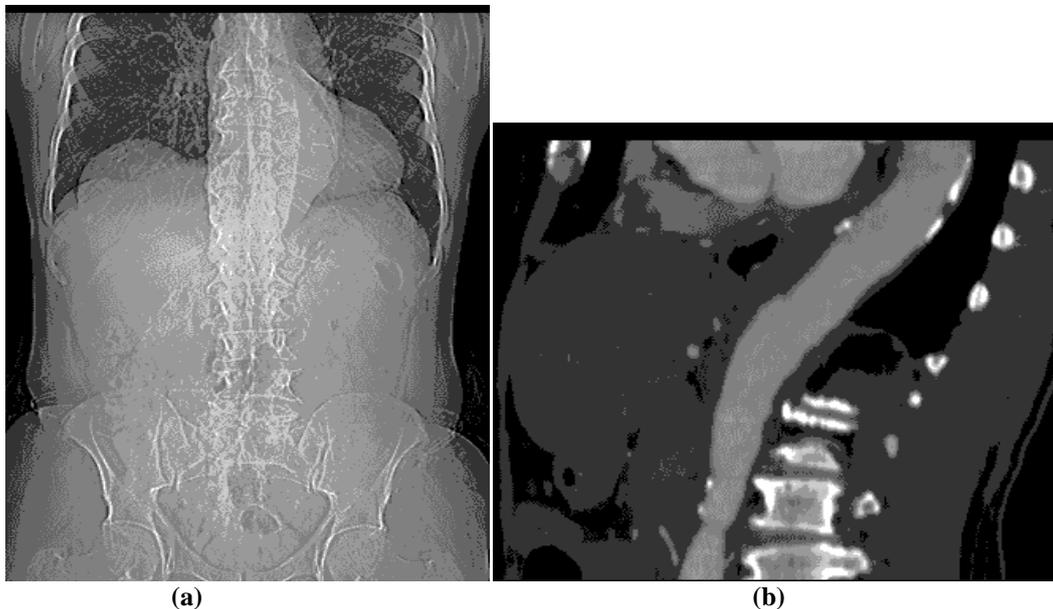


Fig 2: (a) & (b): CT scanogram and reformatted images show fusiform aneurysm of thoracic part of descending aorta.

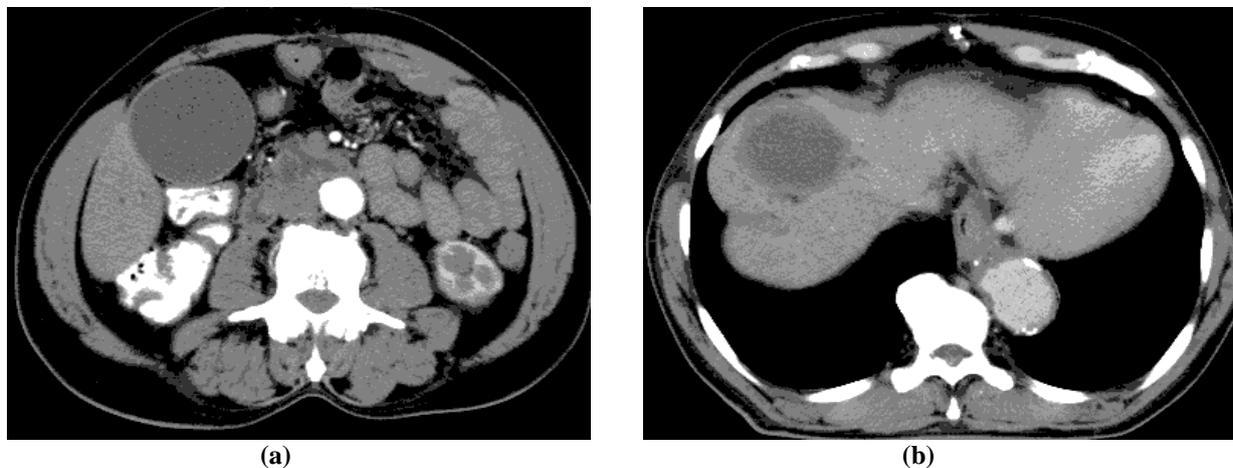


Fig 3: CECT axial: (a) and (b): Ampullary carcinoma and Complicated/ Hydatid Cyst in liver.

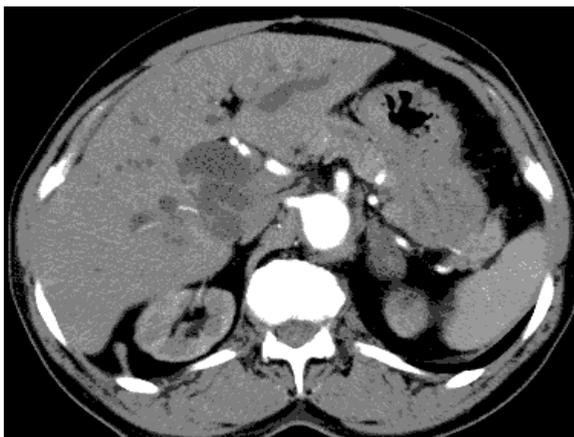


Fig 4: CECT axial: Enhancing lesion noted in left adrenal gland – Adrenal metastasis.



Fig 5: (a) and (b): Plain and CECT Abdomen coronal: Gross IHBR dilatation. Common bile duct and main pancreatic duct are dilated throughout its course and measures ~ 14 mm at porta hepatis.

DISCUSSION:

Ampullary cancer is a rare malignant neoplasm develops in the ampulla of Vater which arises from the tip of the common bile duct, where it passes through the wall of the duodenum and ampullary papilla [1]. The common bile duct merges with the pancreatic duct of Wirsung to form a common channel that exits through the ampulla into the duodenum. The most distal portion of the common bile duct forms the ampulla of Vater and is surrounded by the sphincter of Oddi, which spirals upward around the terminal portion of the duct [2]. According to the American Cancer Society (ACS), nearly everyone who has ampullary cancer has jaundice. Because ampullary cancer causes such a noticeable symptom, it is often found at an earlier stage than pancreatic cancer [1]. Jaundice is caused by an excess of bilirubin in the blood and body tissue. The

tumor in ampulla of Vater can block the common bile duct, preventing bile from reaching the intestines and causing a build-up of bilirubin.

Periampullary carcinoma is usually associated with adenocarcinoma arising from adenoma, papillary adenocarcinoma, mucinous adenocarcinoma, and signet ring cell carcinoma. Rarely are they associated with adenocarcinoma of duodenum and Cholangiocarcinoma [3].

The most common sites to metastases are liver and the lymph nodes. Lymph node metastases are seen in half of patients with periampullary carcinoma [4]. Pericanalicular lymph nodes are the first to be involved followed by the lymph node along the superior mesenteric, gastroduodenal, common hepatic, and

splenic arteries, as well as the celiac trunk. Involvement of Perineural, vascular, and lymphatic invasion are associated with a poor prognosis. In advanced cases, lung metastasis also may occur [3].

Adrenal metastasis in ampullary carcinoma is unusual and rare gastrointestinal malignancy with other associations as discussed. Previously only one other case of adrenal metastasis was reported in periampullary carcinoma [5]. The present of thoracic aortic aneurysm and hydatid cyst in liver were incidental findings associated with the present malignancy.

Differential diagnosis:

- Duodenal Adenocarcinoma
- Pancreatic Carcinoma With Duodenal Invasion

Duodenal Adenocarcinoma

Adenocarcinoma is the most common primary malignancy of the small bowel, with 50% occurring in the duodenum, especially near the ampulla [6]. These tumors typically manifest as an annular narrowing with abrupt concentric or irregular overhanging edges, a discrete nodular mass, or an ulcerated lesion [7]. On CT, these lesions are usually appearing hypovascular and involve only a short segment of bowel, with gradual narrowing possibly leading to obstruction. CT is helpful in the detection of the primary disease, local invasion, potential complications such as obstruction, and nodal or distant metastases [6].

Pancreatic Carcinoma with Duodenal Invasion

Pancreatic carcinomas have the highest likelihood of larger size at detection, poor histologic differentiation, and lymphatic and perineural extension among periampullary carcinomas [8]. CT has been established as the primary method for diagnosing and staging pancreatic cancers. On contrast-enhanced CT, tumors present as heterogeneous poorly enhancing lesions that are relatively hypovascular to pancreatic parenchyma.

CONCLUSION:

Ampullary carcinoma is the second most common cancer of the ampullary area after pancreatic carcinoma and metastasizes mostly intra-abdominally and to the liver. Extra-abdominal metastases are less frequent. In this case report we describe the case of a patient with Ampullary carcinoma who developed adrenal metastasis with gross IHBR dilatation, CBD and MPD dilatation, and coexistence of other conditions like thoracic aorta aneurysm and liver hydatid in the same time. Adrenal metastasis in ampullary carcinoma is unusual and rare gastrointestinal malignancy with other associations as discussed. Previously only one other case of adrenal metastasis was reported in

periampullary carcinoma which has been described in English literature.

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