Duodenal carcinoid - A very rare cause of gastric outlet obstruction (Duodenography-old is gold), with review of literature.

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Citation

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Abstract

Acquired gastric outlet obstruction (GOO) by a duodenal carcinoid is a very rare condition. We report an unusual case of 21 year old female who presented with epigastric fullness and postprandial upper abdominal distention of two and a half year duration and recurrent non bilious vomiting and progressive weight loss of two months duration. Confirmatory preoperative diagnosis was not available. The case was diagnosed as carcinoid postoperatively after pathologic examination. We are reporting this case because of its rarity and with excellent outcome if diagnosed and managed properly. Even on extensive search of medical literature we are not able to find report of such a lesion causing features of gastric outlet obstruction.

CASE REPORT

A 21-year old female was referred to our teritiary care hospital from a peripheral centre for management of epigastric fullness and postprandial upper abdominal distention of two and a half year duration and recurrent non bilious vomiting and progressive weight loss of two months duration. Her past surgical history consisted of appendicectomy performed two years back for right iliac fossa pain.

On admission, the girl was emaciated, weighed 45 Kgs. She had gross pallor. Abdominal examination revealed epigastric distention and a tympanic mass in the epigastric area was appreciated. Bowel sounds were sluggish and per-rectal examination revealed nothing significant. Her herniated orifices were normal.

Her blood investigations were unremarkable except for a total leukocyte count count (TLC) of $17.06x10^6$. Her serum amylase level was normal. Plain X-ray abdomen showed features of a distended stomach. Ultrasonography (USG) abdomen was normal. Upper GI endoscopy was done which was in favour of a duodenal tumour (in the second part, likely a gastro intestinal stromal tumour). Barium study (duodenography) of upper GI tract revealed a well circumscribed filling defect in contrast filled duodenum. The patient underwent elective laparotomy. Duodenotomy (in second part of duodenum) and sub mucous resection of duodenal tumour (located on posteromedial wall) was done. The specimen was sent for pathologic examination which showed the tumour to be carcinoid. The patient became symptom free and was discharged on 7^{th} postoperative day. At 4½ month follow up, she is free of any symptom and thriving well. The patient has also undergone upper GI endoscopy and USG abdomen twice since then, and shows no recurrence till date.

Figure 1

FIGURES: Barium study (duodenography) showing 1.81 x 1.63 cm filling defect in the second part of duodenum (on posteromedial wall)

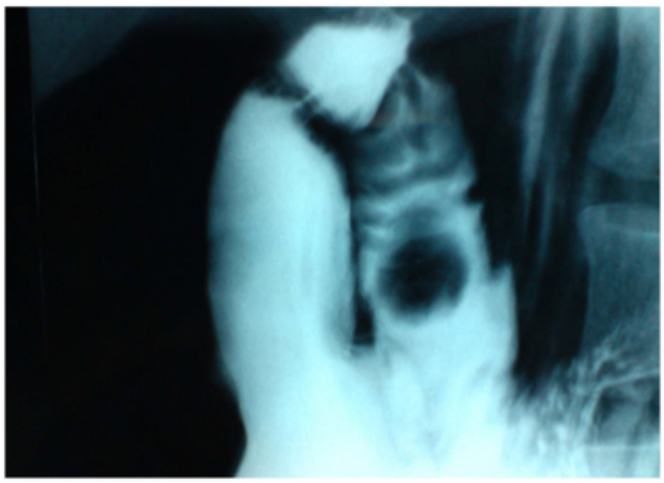


Figure 2

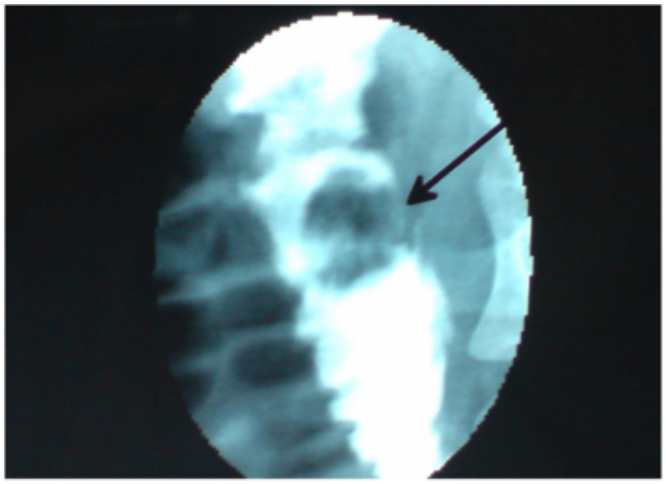


Figure 3

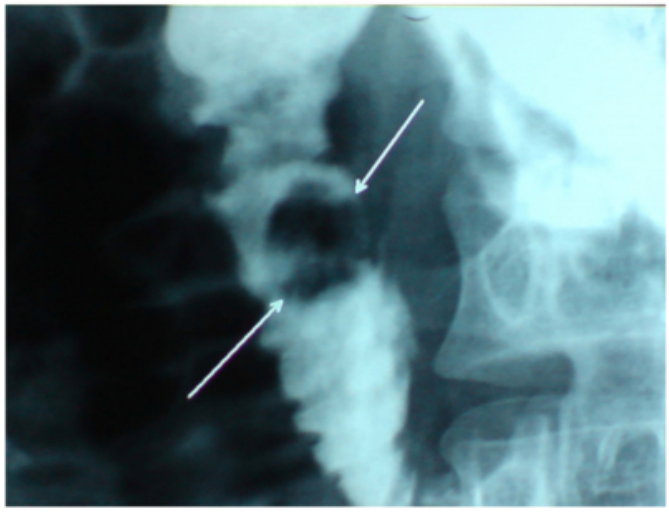


Figure 4

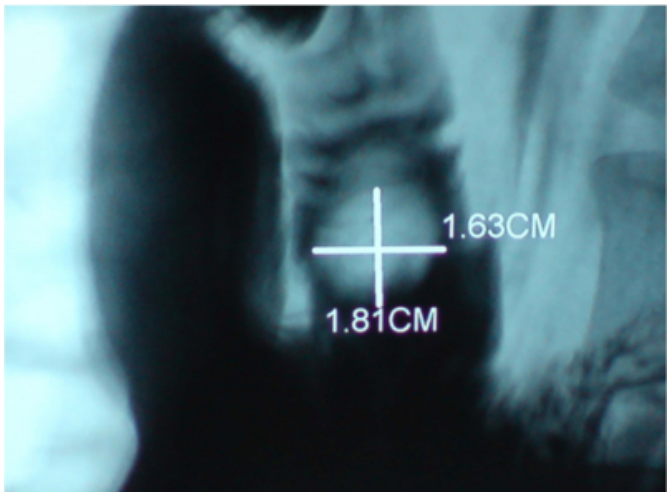
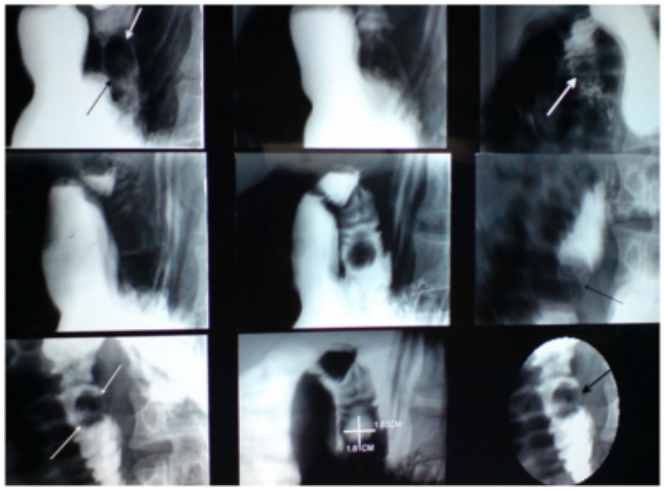


Figure 5



DISCUSSION

Carcinoids are well-differentiated endocrine neoplasms that belong to a diverse group of tumors that arise from cells of the diffuse endocrine system. A wide variety of specialized endocrine cells that populate the gastrointestinal mucosa and submucosa give rise to carcinoids. Consequently, carcinoids may occur throughout the gastrointestinal tract and produce a variety of hormones and protein products that are associated with specific clinical symptoms. Biologic behavior of carcinoids varies by site and cell type, but all gastrointestinal carcinoids are considered to have malignant potential. They may produce specific syndromes such as Zollinger-Ellison syndrome, or they may occur in association with inherited syndromes such as multiple endocrine neoplasia type 1 or neurofibromatosis type 1. Metastatic carcinoids may produce carcinoid syndrome. Carcinoid tumors have been found in a wide range of organs. They most frequently involve the gastrointestinal tract; however, duodenal carcinoid tumours are rare[1]. Here, we report a case where carcinoid involved the rare location – duodenum, causing features of gastric outlet obstruction. Carcinoid tumours arise from neuroendocrine cells that line the tract[1]. Carcinoid tumours most frequently involve the gastrointestinal tract and the bronchopulmonary system[1]. In a report by USA, carcinoid tumours happen most frequently in the gastrointestinal tract (67.5%) and then the respiratory tract (25.3%). Small bowel (41.8%) is the most frequent site of the gastrointestinal carcinoid tumours, followed by the rectum (27.4%) and the appendix (24.1%)[1]. The frequency of duodenal carcinoid tumours is about 2-4% in all carcinoid tumours[1]. There has been one case report of a duodenal carcinoid involving accessory papilla of the pancreas

divisum[2]. Up to 26% of patients may have associated Von Recklinghausen disease[3]. Incidence of carcinoid tumours in the United States has been about 1-2 cases per 100 000 people[4,5]. Small intestinal carcinoids often have an aggressive biologic behavior and, as such, patients frequently have metastases to regional lymph nodes and the liver at initial presentation. In contrast, carcinoids of the appendix and rectum are commonly discovered incidentally as small lesions that are unassociated with clinical evidence of hormone production and have a more indolent clinical course. A study, which compares the duodenal carcinoid tumours with that in other organs, showed an average age of 55.9 years, a relative small average tumour size of 17.7 mm, a relative low metastasis rate of 27.4%, a high postoperative survival rate of 83.3%, and a reasonable incidence of the carcinoid syndrome of 3.1%[6]. They are usually small, solitary lesions confined to the duodenal mucosa had a 5-year death rate of 5.3%[7]. Unlike carcinoids of the jejunum and ileum, duodenal carcinoids have a location advantage and are often discovered endoscopically[7]. Upper GI endoscopy was done in our patient which was in favour of a duodenal tumour (second part). There are fewer than 80 reported cases of carcinoid tumours of the ampulla vater in the literature[8,9]. The advantageous location makes it easier to discover the tumour in comparison to carcinoid tumours, which are located more distally in the small intestine. Carcinoid tumours are associated with an increased incidence of secondary primary malignancies, especially adenocarcinoma[10]. The large intestine is the most common site for synchronous primary malignancies to occur with gastrointestinal carcinoid tumours[11,12]. Duodenal carcinoid have been seen to be associated with gastric leiomyo-blastoma[13] and adenocarcinoma of gastro-esophageal junction[10] (synchronous primary tumours). There have been three case reports describing the development of isolated carcinoid tumours in patients with myelofibrosis[14,15]. Carcinoids have an intact overlying mucosa which may explain a high rate of false negative biopsies[16, 17]. CT scan and MRI have a low sensitivity for the primary lesion[18].

Gastric outlet obstruction caused by a duodenal carcinoid is a very rare entity reported in literature. Our patient had epigastric fullness and postprandial upper abdominal distention of two and a half year duration and recurrent non bilious vomiting and progressive weight loss of two months duration. She had symptoms of obstruction only. No symptoms related to serotonin secreation were present. Surgery, if possible, is the only curative therapy. Duodenotomy (in second part of duodenum) and sub mucous resection of duodenal tumour was done in our patient. Choosing the best treatment option for ampullary carcinoids could be challenging. Although tumor size is not a good prognostic predictor[16,19], most experts still recommend either a Whipple or a pylorus preserving pancreatico-duodenectomy (PPPD) for tumors over 2 cm in size[20]. Metastatic disease has been found in 48% and 40% of the patients with tumor sizes of more than 2 cm and less than 2 cm, respectively[21]. Therefore, some suggest Whipple pancreatico-duodenectomy for all size tumours. On the other hand, long-term survival has been achieved with local resection only[16,22]. In a study of 90 patients with ampullary carcinoids, 52 had a PPPD (majority > 2 cm), while 22 underwent a local resection (majority < 2 cm). Post-operative mortality was 3/52 in the PPPD group compared to 0/22 in the group with local resection[23]. So, less radical approaches should be considered in highly differentiated, slow growing tumours. In high risk surgical patients with a small, non-metastatic tumour, local excision seems to be a reasonable operative choice. An endoscopic ampullectomy which seems to be an effective treatment in the management of ampullary adenomas, could also be a viable option in selected patients with tumours in situ (Tis)[24]. If the tumour has metastasized (most commonly, to the liver) and considered incurable, there are some promising treatment modalities, such as radiolabeled octreotide, which arrests the growth of the tumors and prolongs survival in patients. Chemotherapy is of little benefit and is generally not indicated. Octreotide (a somatostatin analogue) may decrease the secretory activity of the carcinoid. They have a very slow growth rate compared to most malignant tumours. Lastly, it is emphasized that such an atypical case presentation may keep the surgeon in a diagnostic dilemma. The surgeon should keep in his mind, such a possibility while dealing with a case of gastric outlet obstruction, although, possibility of getting such a diagnosis is very rare.

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