

Case Report

A case report of duodenal carcinoid tumor causing gastric outlet obstruction

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Received: 23 April 2021

Revised: 22 May 2021

Accepted: 24 May 2021

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ABSTRACT

Carcinoids are tumours of neuroendocrine origin. Commonly found in gastrointestinal and respiratory tracts, however, duodenal carcinoids among them are comparatively rare. Duodenal carcinoids presenting as acute gastric outlet obstruction are even rarer. Clinically difficult to diagnose due to their non-specific presentation. Endoscopic ultrasound (EUS), computed tomography (CT) and immunohistochemistry (IHC) for markers like chromogranin A, neuron specific enolase (NSE), synaptophysin helps in making a definitive diagnosis. Management is influenced by multiple factors like size, site, metastases and regional lymph node involvement. Here is a case report of solitary duodenal carcinoid complicated with acute gastric outlet obstruction. A middle-aged female with chronic history of intractable dyspepsia, bloating and occasional vomiting, on thorough evaluation with upper GI endoscopy, EUS guided biopsy, CT scan and histopathological examination was diagnosed of a solitary 2.2×1.2×1.6 cm sized duodenal carcinoid tumour. Patient had no past, family or genetic history supporting the diagnosis. Patient presented with features of acute gastric outlet obstruction 1 week after the diagnosis which required a distal gastrectomy with resection of first part of duodenum followed by a Roux-en-Y gastrojejunostomy with an uneventful 6 month follow up. As the size in this case was more than 2 cm with AJCC staging of T2N0M0, distal gastrectomy with Roux-en-Y gastrojejunostomy was done to ensure an R0 resection and to relieve the gastric outlet obstruction. Duodenal carcinoids presenting as gastric outlet obstruction are not common. Early management is essential to prevent complications like gastric outlet obstruction despite of indolent course of the disease.

Keywords: Duodenal carcinoid, Neuroendocrine, Chromogranin A, Synaptophysin, Gastric outlet obstruction

INTRODUCTION

Carcinoid tumours originate from the cells of neuroendocrine origin which are dispersed among different organ systems like the Gastrointestinal tract, lungs, bronchi, ovaries and the testicles. They were first described by Otto Lubarsch in 1888. Oberndorfer coined the term 'Karzenoide' in 1907, which meant carcinoma like small bowel tumour which behaved in a benign fashion.¹ They have an indolent course and are usually more benign than the tumours of epithelial origin of the

same anatomical sites. Depending on the cell of origin and site, the biological aggressiveness exhibited by them is variable and unpredictable.

Duodenal carcinoids are comparatively rare amongst the carcinoids of the GIT.² They are slow growing with low metastatic potential compared to their counterparts in the rest of the GIT. Usually patients are asymptomatic, incidentally diagnosed on gastroduodenoscopy performed for other causes or they might present with non-specific abdominal symptoms. Upper GI endoscopy with EUS not

only aids in procuring the biopsy, but also helps in evaluation of the depth of invasion and locoregional spread. Contrast enhanced CT is a prerequisite to diagnose distant metastases, which are more common with primaries presenting with carcinoid syndrome. IHC with special stains for chromogranin A, NSE, synaptophysin makes a more specific diagnosis of carcinoid tumour.

We report a case of duodenal carcinoid complicated with acute gastric outlet obstruction.

CASE REPORT

A middle-aged female who presented with initial features of non-specific abdominal symptoms. On thorough evaluation with EUS, CT and silver staining immunohistochemical techniques, a 2.2 cm duodenal carcinoid tumour limited to the submucosa was diagnosed, surprisingly with no locoregional metastases considering the large size for the given location. Despite of non-aggressive course of the disease, it later progressed to acute gastric outlet obstruction necessitating a distal gastrectomy with resection of first part of duodenum followed by a Roux-en-Y gastrojejunostomy.

A 55-years-old middle-aged Indian female, office worker, presented to with complaints of abdominal distension, pain in abdomen and recurrent vomiting for 2 days. Abdominal distension was of acute onset, associated with mild, non-radiating, colicky pain in epigastrium and umbilical region. Recurrent episodes of non-projectile, non-bilious vomiting immediately after food intake were also present. Patient had intractable dyspepsia, bloating, occasional vomiting and repeated OPD visits for her complaints since past 6 months which were not reduced on medical therapy. She had no comorbidities, no history of substance abuse or any kind of relevant family/surgical history. Clinically, the patient had only a mildly distended abdomen.

Gastroduodenoscopy was performed 1 week before the current presentation, which revealed an abnormal sessile mucosal elevation in the first part of the duodenum in the anterior aspect. Endoscopic ultrasound revealed a well-defined, hypoechoic mass of 2.2 cm size in the anterior wall of the first part of the duodenum in the deep submucosa, adjacent to muscularis propria without any involvement of muscularis or local lymphadenopathy. Adequate Endoscopic biopsy of the lesion was obtained. Histopathological examination of the same with haematoxylin and eosin staining demonstrated a well circumscribed lesion in submucosa with monomorphic islands of small cells with round nuclei surrounded by scarce cytoplasm.

A provisional diagnosis of carcinoid tumour was made and the same was confirmed after positive reaction to markers like neuron specific enolase (Figure 1), chromogranin A (Figure 2) and synaptophysin (Figure 3). Contrast enhanced computed tomography (Figure 4) of the abdomen demonstrated a solitary neoplasm measuring

2.2×1.2×1.6 cm in first part of duodenum with contrast enhancement in initial venous phase with no distant metastases, lymphadenopathy and desmoplastic reaction.

Patient presented now with acute onset abdominal distension, pain and non-bilious vomiting i.e.; features of gastric outlet obstruction. Cell counts and serum biochemistries were unremarkable. Urinary 5 HIAA levels were not elevated (<6 mg/24 hr) and patient had no symptoms suggestive of carcinoid syndrome. After diagnosis of the lesion with radiological, histopathology and immunohistochemistry, midline laparotomy with distal gastrectomy and resection of first part of duodenum with Roux-en-Y gastrojejunostomy was performed as the tumour size was more than 2 cm with AJCC staging of T2N0M0, limited to the submucosa and had no locoregional spread. Patient did not require any kind of adjuvant therapy. Histopathological examination of the specimen confirmed a carcinoid tumour of the first part of the duodenum with neuron specific enolase, chromogranin A and synaptophysin positivity.

Post-operative period was uneventful and the patient has no new onset complaints or recurrence on 6 months follow up EUS.

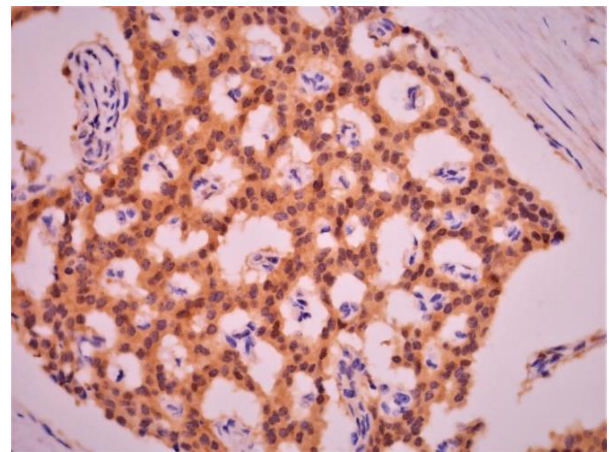


Figure 1: Neuron specific enolase.

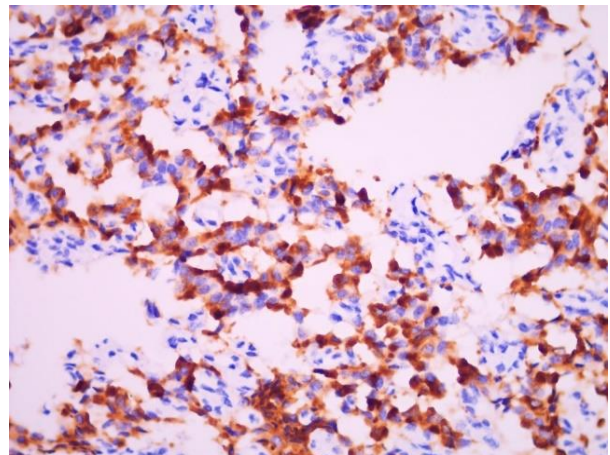


Figure 2: Chromogranin A.

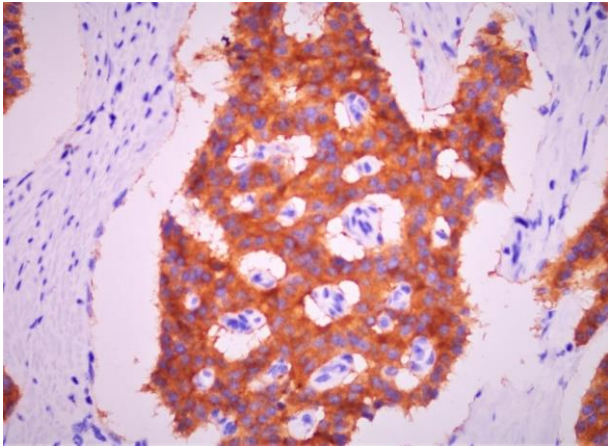


Figure 3: Synaptophysin.



Figure 4: Coronal view of contrast enhanced CT of the abdomen demonstrating a hypointense tumour in first part of duodenum.

DISCUSSION

Carcinoid tumours are diverse group of neoplasms originating from variety of neuroendocrine cells which occur most commonly in the gastrointestinal tract. They are also seen in the lungs, bronchi, ovaries and testicles. They have an indolent course and are usually more benign than the tumours of epithelial origin of the same anatomical sites.^{3,4} Depending on the cell of origin and site, the biological aggressiveness exhibited by them is variable and unpredictable. Though, commonly found in appendix, terminal ileum, stomach and rectum, duodenal carcinoids are rare entities.

Primary duodenal carcinoids constitute less than 2% to overall incidence of gastrointestinal carcinoids with an annual incidence of 0.07/1,00,000 patients, with a slightly higher male preponderance.^{2,5} With advent of routine

upper GI endoscopy, the incidence of duodenal carcinoid diagnosis is increasing. Compared to their ileal counterparts, occurrence of the carcinoid syndrome (seen in 4% of cases) and metastatic potential is less common in duodenal carcinoids. Tumours originating in the ampullary region possess a higher metastatic potential compared to non-ampullary carcinoids, irrespective of their size. The WHO classifies carcinoids into well differentiated endocrine tumours, well differentiated endocrine carcinomas and poorly differentiated endocrine carcinomas.^{6,7} They are also classified into typical and atypical types based on mitotic figures and presence of necrosis.⁸

Secretion of bioactive amines and peptides like serotonin, gastrin, somatostatin, glucagon etc is a hallmark of these tumours depending on the cell of origin. However, chromogranin A, neuron specific enolase, synaptophysin are more common and specific.

Patients are usually asymptomatic; however, epigastric pain, dyspepsia, vomiting, bleeding and weight loss can be present.^{9,10} Preoperative diagnosis is made with serum chemistries, immunohistochemistry and radiology. Gastroduodenoscopy demonstrates intraluminal irregular/smooth elevations in the duodenal mucosa.

EUS plays a vital role in visualizing the depth of duodenal wall invasion, local lymph node metastasis and size of the tumour.^{11,12} Contrast enhanced CT is helpful in demonstrating hepatic and distant metastasis and staging the tumours. As most of these tumours exhibit high somatostatin receptor density, radiolabelled octreotide (octreoscan) scan is a recent modality of investigation to detect the primary neoplasm and the distant metastases.

The management of duodenal carcinoids depends on the size of the tumour, involvement of lymph nodes, differentiation and metastases. Invasion of the muscular layer, size >2 cm and presence of >2 mitotic figures/hpf are considered to be independent risk factors for metastases as per study conducted by Burke et al.¹³ Tumours <1 cm size which are non-ampullary, with no muscularis involvement and no regional lymphadenopathy are resected endoscopically (EMR). Management of tumours 1-2 cm size is controversial.

Trans-duodenal full thickness resection is the treatment modality for tumours of 1-2cm size and when the involvement is beyond submucosa.^{4,14,15} Surgical resection with re-anastomosis is done for tumours >2cm size and when the mitotic index is >2/hpf with or without regional lymphadenectomy based on the CT findings.⁸ However, pancreaticoduodenectomy may be required for ampullary and periampullary carcinoids.

Our patient had minimal non-specific symptoms for 6 months despite of 2.2 cm duodenal carcinoid, later complicated with acute gastric outlet obstruction, 1 week after the diagnosis. As the tumour was more than 2.2 cm

in size, resection of the affected segment with re-anastomosis was done. Duodenal carcinoids presenting with features of gastric outlet obstruction are rare. Despite the indolent course of the disease and better prognosis compared to tumours of epithelial origin of the same location, aggressive management is essential to prevent acute complications.

CONCLUSION

Non ampullary duodenal carcinoids presenting as gastric outlet obstruction are rare. Though indolent in course and non-aggressive in behaviour, they can precipitate complications like gastric outlet obstruction. Early diagnosis and treatment will help in prevention of the same.

ACKNOWLEDGEMENTS

Authors would like to thank Department General Surgery, Grant government medical college and JJ group of hospitals, Mumbai, India for valuable support during study.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

REFERENCES

1. Kloppel G. Oberndorfer and his successors: from carcinoid to neuroendocrine carcinoma. *Endocr Pathol.* 2007;18(3):141-4.
2. Nikou GC, Toubanakis C, Moulakakis KG, Pavlatos S, Kosmidis C, Mallas E, et al. Carcinoid tumors of the duodenum and the ampulla of Vater: current diagnostic and therapeutic approach in a series of 8 patients. *Case series. Int J Surg.* 2011;9(3):248-53.
3. Attanoos R, Williams GT. Epithelial and neuroendocrine tumors of the duodenum. *Semin Diagn Pathol.* 1991;8(3):149-62.
4. Soga J. Endocrinocarcinomas (carcinoids and their variants) of the duodenum. An evaluation of 927 cases. *J Exp Clin Cancer Res.* 2003;22(3):349-63.
5. Oberg K. Gastrointestinal carcinoid tumors (gastrointestinal neuroendocrine tumors) and the carcinoid syndrome. *Sleisenger and Fordtran's Gastrointestinal and Liver Disease.* 11th ed. WB Saunders; 2010: 475-490.
6. Nikou GC, Toubanakis C, Moulakakis KG, Pavlatos S, Kosmidis C, Mallas E, et al. Carcinoid tumors of the duodenum and the ampulla of Vater: current diagnostic and therapeutic approach in a series of 8 patients. *Case series. Int J Surg.* 2011;9(3):248-53.
7. Washington MK, Tang LH, Berlin J, Branton PA, Burgart LJ, Carter DK, et al. Protocol for the examination of specimens from patients with neuroendocrine tumors (carcinoid tumors) of the small intestine and ampulla. *Arch Pathol Lab Med.* 2010;134(2):181-6.
8. Waisberg J, Joppert NG, Vasconcellos C, Sartini GH, Miranda LS, Franco MI. Carcinoid tumor of the duodenum: a rare tumor at an unusual site. *Case series from a single institution. Arq Gastroenterol.* 2013;50(1):3-9.
9. Han SL, Cheng J, Zhou HZ, Guo SC, Jia ZR, Wang PF. Surgically treated primary malignant tumor of small bowel: a clinical analysis. *World J Gastroenterol.* 2010;16(12):1527-32.
10. Makhoulf HR, Burke AP, Sobin LH. Carcinoid tumors of the ampulla of Vater: a comparison with duodenal carcinoid tumors. *Cancer.* 1999;85(6):1241-9.
11. Levy AD, Taylor LD, Abbott RM, Sobin LH. Duodenal carcinoids: imaging features with clinical-pathologic comparison. *Radiology.* 2005;237(3):967-72.
12. Matsumoto S, Miyatani H, Yoshida Y, Nokubi M. Duodenal carcinoid tumors: 5 cases treated by endoscopic submucosal dissection. *Gastrointest Endosc.* 2011;74(5):1152-6.
13. Burke AP, Sobin LH, Federspiel BH, Shekitka KM, Helwig EB. Carcinoid tumors of the duodenum. A clinicopathologic study of 99 cases. *Arch Pathol Lab Med.* 1990;114(7):700-4.
14. Tai WP, Yue H. Endoscopic mucosa resection of a duodenum carcinoid tumor of 1.2 cm diameter: a case report. *Med Oncol.* 2009;26(3):319-21.
15. Zyromski NJ, Kendrick ML, Nagorney DM, Grant CS, Donohue JH, Farnell MB, et al. Duodenal carcinoid tumors: how aggressive should we be?. *J Gastrointest Surg.* 2001;5(6):588-93.

Cite this article as: Shaikh A, Sholapur S, Tandur A. A case report of duodenal carcinoid tumor causing gastric outlet obstruction. *Int Surg J* 2021;8:2176-9.