Synchronous Gastrointestinal Tumours Encircling Massively Dilated Pancreatic Duct: Duodenal Net and Proximal Jejunal Gist

Maira Aslam, Mariam Shah, Sana Sayeed and Aroosa Kanwal

Department of Radiology, Shifa International Hospital, Islamabad, Pakistan

ABSTRACT

Neuro-endocrine tumours (NETs) arise from neuro-endocrine cells and can potentially occur in any part of the body, the most common site being the gastrointestinal (GI) tract. GI stromal tumours (GISTs) arise from pacemaker cells and are also considered as common GI tumours. However, the synchronous occurrence of these two tumours is rarely reported.

We hereby describe a case of simultaneous occurrence of duodenal NET and proximal jejunal GIST. A 64-year female presented with a long-standing history of obstructive jaundice and significant weight loss. Radiological imaging revealed an extremely dilated pancreatic duct and large heterogenous masses involving the C loop of duodenum and proximal jejunum. She underwent upper GI endoscopy and biopsy which labelled duodenal mass as well-differentiated NET, grade one and jejunal mass as GIST. Whipple's procedure is in the plan after the improvement of general physical health to bear the surgical burden.

Fortunately, the presence of two distinct low-grade tumours in such close approximation does not change the management plan. Surgical excision is the definite treatment of choice.

Key Words: *Neuroendocrine tumour, Gastrointestinal stromal tumour, Obstructive jaundice.*

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INTRODUCTION

Neuro-endocrine tumours (NET) are heterogenous tumours with an indolent course, particularly the non-functional type. They are typically diagnosed later due to their increased size and mass effect. With the increasing availability of radiological imaging, these masses are identified and managed at an earlier stage. Gastrointestinal stromal tumours (GIST) can arise from any part of the gastrointestinal (GI) tract with the stomach (60%) being more common than the duodenum (5%).[1]

The described cases of synchronous GIST and duodenal NETs are very few in the literature. A few cases that have been reported include patients with a known history of neurofibromatosis (NF).[2] Moreover, a few cases of ampullary NET co-existing with GIST have been described without causing significant obstruction of the pancreatic duct (PD) and common bile duct (CBD). This unique case describes a large duodenal NET causing extreme PD and CBD dilatation. This extreme PD dilatation can be easily misdiagnosed as a primary cystic pancreatic mass.

Correspondence to: Dr. Maira Aslam, Department of Radiology, Shifa International Hospital, Islamabad, Pakistan
E-mail: maira123.ma4@gmail.com

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To the best of our knowledge, this is the first case reported from Pakistan describing duodenal NET and jejunal GIST causing severe dilatation of PD. Our patient had no history of NF, suggesting that such atypical presentation of the otherwise two distinct tumours can occur and that too in patients with no history of apparently well-associated risk factors.

CASE REPORT

A 64-year female patient presented to GI OPD with complaints of abdominal pain, jaundice, vomiting, and weight loss for the last six months. Apart from a prior history of cholecystectomy and bile duct stenting about 10 years back, she had no significant medical or surgical history. On general examination, she was an emaciated lady with yellowish skin and sclera. The abdomen was soft and non-tender with no palpable mass. She was admitted for further work-up and underwent a pancreatic CT dynamic considering the possibility of a pancreatic lesion. CT showed multiple large heterogenous lobulated enhancing masses involving the C loop of the duodenum, with sparing of its first part and involving duodenojejunal (DJ) flexure and the proximal jejunum (Figure 1). The ampulla and periampullary regions were not visualised, likely effaced with massively dilated and obstructed PD and CBD. PD was disproportionately dilated, reaching up to 60 mm in calibre (Figure 2). No normal pancreatic parenchyma was visualised, likely atrophied owing to significant compression. No wall abnormality or intraductal soft tissue mass was present to suggest a primary pancreatic ductal tumour. Apart from a few regional pathological lymph nodes,

there was no local direct extension or distant metastasis. Moreover, chronic endobiliary stents were seen in the left hepatic duct and CBD surrounded by encrustation and calculi.

Laboratory tests showed raised gamma-glutamyl transferase of 662 U/L (normal up to 40 U/L) and alkaline phosphatase levels of 1083 U/L (normal: 40-130 U/L). CA-19-9 was within normal limits at 25 U/ml (normal is up to 34 U/ml).

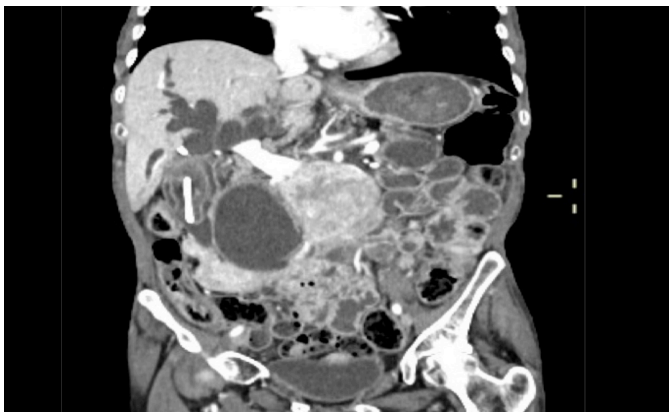

Figure 1: Lobulated heterogeneously enhancing mass involving duodenum, duodenojejunal (DJ) flexure, and proximal jejunum.

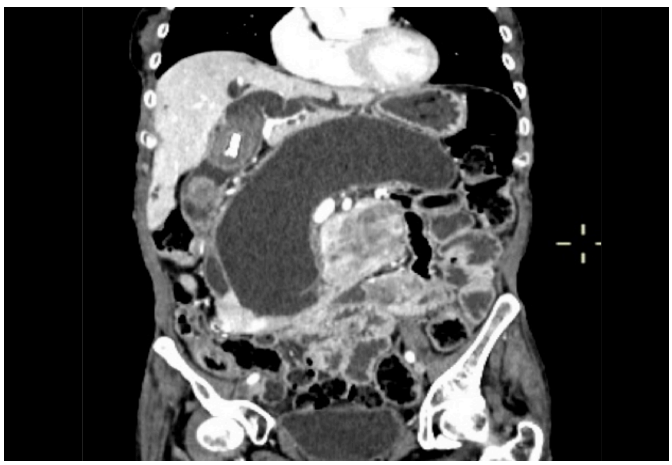

Figure 2: CT coronal reformatted image showing massively dilated pancreatic duct and common bile duct with internal encrusted stent. Bulky heterogeneously enhancing mass conforming the contour of the duodenum and duodenojejunal (DJ) flexure.

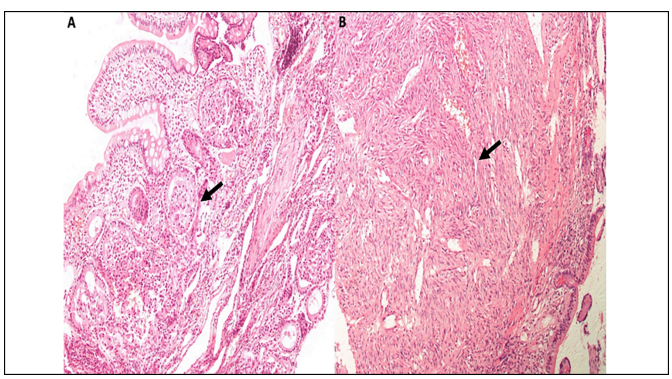

Figure 3: (A) Duodenal biopsy: Well-differentiated neuroendocrine tumour (WD-NET) with a nested and trabecular growth pattern. (H and E stain, ×20). (B) Jejunal mass: Gastrointestinal stromal tumour (GIST) characterised by spindle-shaped cells with eosinophilic cytoplasm and variable cellularity (H and E stain, ×20).

The patient underwent an upper GI endoscopy in which biopsies were taken from ulcerated masses in the second and third parts of the duodenum and proximal jejunum. Histopathology of duodenal masses showed a well-differentiated NET, grade one and that of jejunal mass as GIST (Figure 3 A,B). Duodenal biopsy showed nests of atypical cells having hyperchromatic nuclei and salt/ pepper chromatin. Immunohistochemistry (IHC) showed positive Cam 5.2, *Synaptophysin*, and *Chromogranin* with Ki-67 less than 1%. Jejunal biopsy showed spindle-shaped cells with eosinophilic cytoplasm and variable cellularity. IHC showed positive *DOG1* and *CD117* and negative *ASMA* markers.

After establishing the diagnosis of the case, the therapeutic management was discussed as surgical *vs.* conservative. Endoscopic treatment was not feasible considering the large tumour size analogue. The patient was referred to the hepatobiliary department, and surgical resection / whipple's procedure was planned.

DISCUSSION

NETs compromise about 2 to 8% of GI tumours with G cell tumour being the most common duodenal NET followed by D-cell somatostatinoma.[3]

Ninety percent of D-NETs are non-functional. Most of the NETs are detected incidentally on endoscopy while causing symptoms related to elevated gastrin levels and carcinoid hormones. D-NETs are divided into 3 grades based on WHO classification, with grade one being the most common.[4]

GISTs are the most common tumours of GIT derived from interstitial cells of Cajal initiated by a mutation in tyrosine kinase. Most are detected incidentally, but symptoms depend upon size and location.[5]

Imaging features of these tumours also vary with location typically appearing as hyper-enhancing endoluminal or exophytic masses.

Regarding the literature review, there are few cases available describing the co-existence of duodenal NETs and jejunal GISTs. Most of the cases described had a history of NF1. The present patient had no history of NF1 but showed the co-occurrence of these two tumours.

Marked mass effect without causing frank invasion is also typical of slow-growing tumours, as evident in this case, where masses were well-confined within the epicentre but caused enormous PD dilatation. Duodenal adenocarcinomas are more common than D-NETs, but the imaging features were not typical. Therefore, correct interpretation of imaging findings can accurately predict the grade as well as the type of tumour. Surgical resection is the standard treatment of choice, with Whipple procedure reserved for larger and locally advanced tumours.

The fact that the two distinct low-grade GI tumours can co-exist in close approximation and can cause significant mass effects on surrounding structures should be acknowledged. Documen-

tation of such occurrences will help the primary team for appropriate surgical planning, reducing morbidity, and achieving greater patient satisfaction regarding the diagnosis and treatment.

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PATIENT'S CONSENT:
Informed consent was taken from the patient.

COMPETING INTEREST:
All authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:
MA: Substantial contributions to the conception or design of the work.
MS: Drafting of the work and revising it critically for important intellectual content.
SS: Final approval of the version to be published.
AK: Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy and integrity of any part of the work are appropriately investigated and resolved.
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