

# Giant gastrointestinal stromal tumor of the duodenum mimicking a pancreas head tumor: a case report

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## ABSTRACT

**Objectives:** Duodenal gastrointestinal stromal tumor (GIST) is rarely observed in the gastrointestinal system. It can be confused with pancreas pathologies as it shows an extramural growth pattern. The case is here presented of a 65-year-old male who presented with the complaints of abdominal pain, nausea and vomiting. On contrast abdominal tomography, a centrally hypodense, necrotic mass, 140×115×100 mm in size, was observed in the pancreas head. With a preliminary diagnosis of neuroendocrine tumour/serous cystic neoplasia, a Whipple + partial vena cava resection + primary repair operation was performed. The pathology was reported as duodenal GIST. The patient was discharged with medication and referred to the oncology clinic.

**Keywords:** Duodenum, gastrointestinal stromal tumor, pancreas, Whipple

Gastrointestinal stromal tumor (GIST) originates from the interstitial cells of Cajal and is found in the submucosa of the gastrointestinal system and the myenteric plexus. There are 3 histological types: spindle cell type, which is the most common, epithelioid type, and mixed type [1]. GISTs are specific mesenchymal neoplasia, which can be observed in the digestive system from the mouth to the anus. They are generally seen in the gastric (60%-70%), small intestine (30%) and colon-rectum (10%), and are only seen in the duodenum at the rate of 3%-5%. Duodenal stromal tumors are rarely seen. Tumors with an extramural growth pattern which can clinically and radiologically mimic pancreas head cancer are extremely rare [2]. The case is here presented of a patient with a spindle cell duodenal GIST which appeared as a mass on the pancreas head.

## CASE PRESENTATION

A 65-year-old male presented at our clinic with complaints of abdominal pain, nausea and vomiting. On physical examination a palpable mass and associated sensitivity was determined in the epigastric region. The laboratory values and tumor markers were normal. Triphasic abdominal computed tomography (CT) with intravenous contrast was applied to the patient. A centrally hypodense, necrotic, mass lesion was observed showing exotic development at the level of the pancreas uncinata process, measuring approximately 140×115×100 mm, which showed significant peripheral contrast in the arterial phase after contrast injection, a relative decrease in contrast in the venous phase, and washout in the late phase. The mass lesion was surrounding the aorta from the anterolateral at ap-

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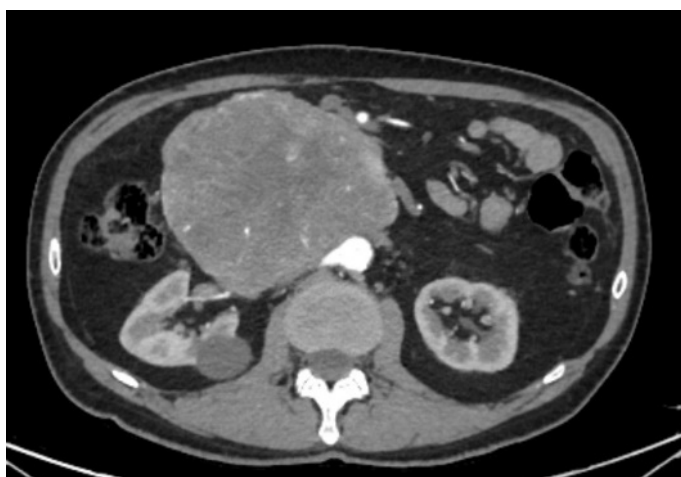
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**Fig. 1.** Triphasic abdominal computed tomography (CT).



**Fig. 2.** Whipple procedure.

proximately 45°. The mass was compressing the vena cava inferior and right ureter. The superior mesenteric artery (SMA) was bowed and pushed towards the anterior by the mass lesion. In the preliminary diagnosis, neuroendocrine tumour showing exotic development and serous cystic neoplasia of the pancreas were first considered (Fig 1). Endoscopic Ultrasonography (EUS) and biopsy was not performed. Because GIST was not considered in the first stage and an operable mass was observed in the head of the pancreas. A Whipple procedure and partial vena cava resection + primary suture repair were performed (Fig. 2). As a result of pathology examination, the mass was reported

as a duodenal GIST, 12 cm in diameter, formed of spindle cells, with mitosis ratio 9/5 mm<sup>2</sup>, DOG1: +++, CD34:-, and Desmin Ki67: >10% high risk. Six lymph nodes dissected from around the pancreas and small intestine were reactive. No complications developed postoperatively and the patient was discharged on day 12 with medication, and was referred to the oncology clinic. Treatment of imatinib was started by the oncology department. On the follow-up tomography at postoperative 18 months, irregular tissue of 25×15 mm was observed adjacent to the SMV. No involvement was observed on PET-CT. The patient was seen by the oncology council and it was decided to continue follow up with a doubling of the imatinib dose.

## DISCUSSION

The first definitive immunohistochemical diagnosis of GIST was made by showing over-expression in KIT (CD117) marker. Nowadays, almost 100% of diagnoses are made from CD117 and DOG 1 positivity. There are known correlations between CD34 and the tumour region, and between CD117 and tumour morphology [3].

The annual incidence of GIST is approximately 10 in 1 million. Although there are a greater number of asymptomatic GIST, determination is difficult. In autopsy series, asymptomatic GISTs < 1 cm have been observed at the rate of 25%. Symptomatic GISTs mostly manifest with dyspepsia, bleeding and abdominal mass [4]. The current patient had dyspeptic complaints and there was an abdominal mass.

Duodenal GIST is uncommon but is mostly located in the second section of the duodenum, followed by the third, fourth, and first sections. In many duodenal GISTs, giant ulcers can be observed in the mucosa due to the pressure of the submucosal mass [5]. Endoscopy, tomography, or MRI can be used in the diagnosis of duodenal GIST. Giant GISTs show a heterogeneous structure on tomography. Together with areas of cystic degeneration, there is hemorrhage and necrosis in the centre. This can therefore lead to incorrect evaluation of giant duodenal GIST as a mass on the pancreas head [6]. Especially in the arterial phase or late venous phase, GIST can be seen as a hypervascular exophytic or endophytic mass [7]. In the current case, there was central necrosis in a giant mass com-

pressing the vena cava and bowing the SMA. In the arterial phase, the mass showed significant peripheral contrast and in the late venous phase, there was washout. In the initial radiological diagnosis, it was thought to be a neuroendocrine tumour (NET) of the pancreas or serous adenocarcinoma.

For duodenal GISTs, just as for other GISTs, the only curative treatment is surgery. Optimal surgery is the removal of the mass with clean surgical margins to include invaded organs. There is no lymph node metastasis in GISTs. They are well-encapsulated tumours showing a submucosal growth pattern. Local excision or duodenectomy is associated with long-term disease-free survival. However, when necessary, pancreatoduodenectomy is applied at rates of 20%-86% [7]. In the current case, a pancreatoduodenectomy operation was performed.

Curative surgery applied with complete tumour resection increases survival. If negative surgical margins are obtained, recurrence can be observed. Pidhorecky *et al* reported the development of recurrence at 76% in the stomach and 64% in the small intestine despite clean surgical margins [8]. On the follow-up tomography of the current case at postoperative 18 months, irregular tissue of 25×15mm was observed adjacent to the SMV. No involvement was observed on PET-CT. The patient was seen by the oncology council and it was decided to continue follow up with a doubling of the imatinib dose.

## CONCLUSION

In conclusion, duodenal GISTs can mimic pancreas head masses because of the extramural growth pattern. This must not be ignored in the differential diagnosis of tumours located in the duodenum and pancreas.

### Authors' Contribution

Study Conception: EA, MAÜ; Study Design: EA, MAÜ; Supervision: OA; Funding: N/A; Materials: EA; Data Collection and/or Processing: OA; Statistical

Analysis and/or Data Interpretation: MAÜ, EBB; Literature Review: EBB; Manuscript Preparation: EA, MAÜ and Critical Review: OA, EBB.

### Informed Consent

Written informed consent was obtained from the patient for publication of this case and any accompanying images or data.

### Conflict of interest

The authors disclosed no conflict of interest during the preparation or publication of this manuscript.

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