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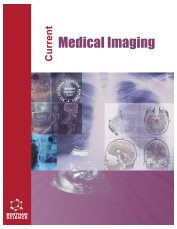

CASE REPORT

A Fistulized Giant Duodenal Stromal Tumor in a Young Patient: A Case Report With Literature Review for Tomographic Diagnosis

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Abstract:

Background:

Duodenal gastrointestinal stromal tumors (GISTs) are rare tumors of the gastrointestinal tract. It should be considered in the differential diagnosis of periampullary region pathologies.

Case Report:

A 24-year-old male patient applied to the general surgery department with the complaint of long-standing abdominal pain, nausea and vomiting after meals, and 8-10 kg weight loss in 1 month. Three-phase dynamic abdominopelvic CT showed that the 1st and the 2nd segments of the duodenum were dilated. At this level, a peripherally intensely contrasted heterogeneous mass lesion, 91x70x46 mm in size, was observed. There was oral contrast and air values in the center of the mass. A fistulized mass connected with the duodenal wall was considered in the differential diagnosis. In the surgical exploration, a soft, vascularized mass fistulized to the 2nd segment of the duodenum was observed. Pathological diagnosis was reported as GIST.

Conclusion:

GISTs arise from the precursors of Cajal Interstitial cells of the gastrointestinal tract. Contrast-enhanced CT is the preferred diagnostic method for staging, risk stratification, and follow-up. We presented a young case with a giant duodenal GIST and discussed differential diagnosis and some diagnostic properties.

Keywords: Gastrointestinal stromal tumor, Case report, Duodenum, Fistula, Tomography, Diagnosis.

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1. INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are the most common non-epithelial tumors of the gastrointestinal tract [1]. They are rare and constitute approximately 1-2% of gastrointestinal neoplasms. They are mostly located in the stomach (60-70%), small intestine (20-30%), colon, and rectum (10%), and less frequently in the esophagus, appendix, anus, and retroperitoneum. Only 1-5% of GISTs occur in the duodenum [2], and the second segment is the most common site of involvement.

Duodenal GISTs have different clinical presentations. Small lesions are usually asymptomatic and detected incidentally, but big lesions may present with severe symptoms such as abdominal pain or discomfort, gastrointestinal bleeding, or bowel obstruction. Also, ulceration of the intestinal wall may occur [3], especially in those with large GISTs. Some prognostic factors have been defined, which include the long diameter of the tumor, the number of mitoses, anatomical localization, and oncogene mutation in tyrosine kinase receptor (KIT) [4, 5].

We present the computed tomography (CT) findings of a large duodenal GIST in a young male and a literature review. The lesion had a giant diverticulum-like appearance and fistulized to the 2nd part of the duodenum.

2. CASE REPORT

A 24-year-old male patient applied to the general surgery outpatient clinic with the complaint of abdominal pain, nausea, vomiting after meals, and 8-10 kg weight loss in 1 month. He had a history of gastric ulcer.

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Three-phase dynamic abdominopelvic CT with the administration of an oral contrast agent was performed with the preliminary diagnoses of duodenal ulcer and diverticulitis. The 1st and the 2nd segments of the duodenum were dilated, and at this level, a peripherally intensely contrasted heterogeneous mass lesion, 91x70x46 mm in size, was observed (Figs. **1**-**4**). There were oral contrast substance and air values in the center of the mass. The border between the duodenal wall and the fat plane was indistinct. The mass extended from the duodenal wall to the inferolateral abdomen region adjacent to the abdominal wall and displaced the ileocecal colon medially.

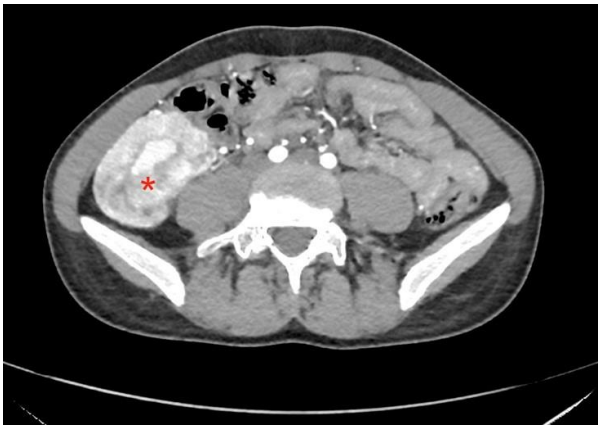

Fig. (1). Duodenal GIST, arterial phase.

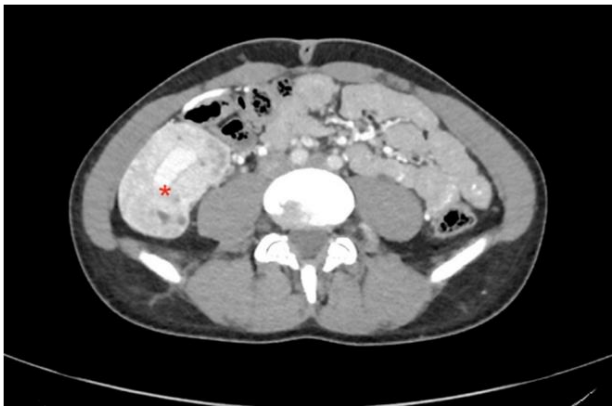

Fig. (2). Duodenal GIST, portal phase.

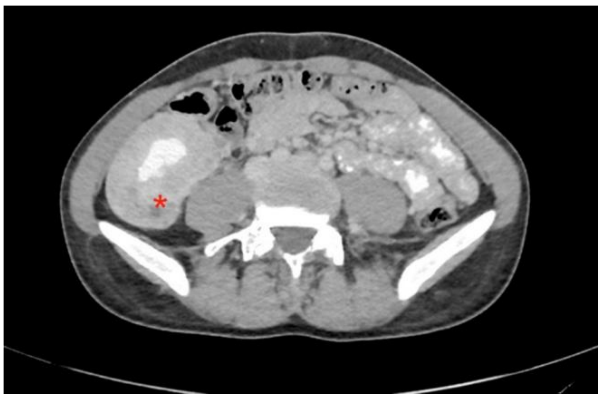

Fig. (3). Duodenal GIST, late phase.

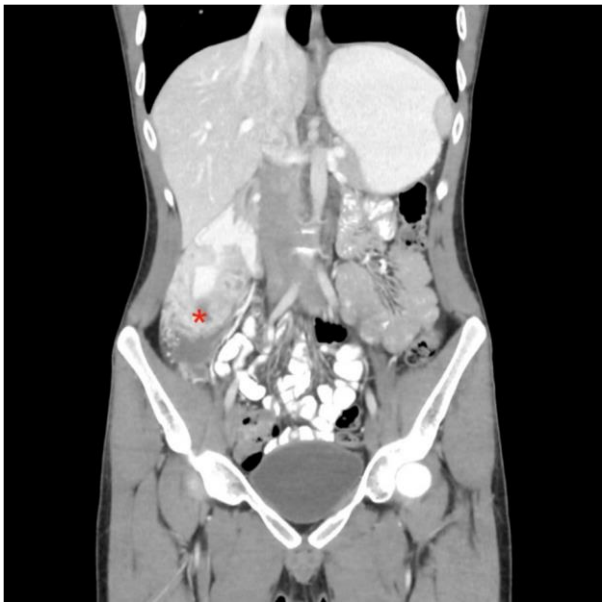

Fig. (4). Duodenal GIST, coronal plane.

Differential diagnoses should include duodenal adenocarcinoma and duodenal lymphoma. Despite the findings of a fast-growing mass (*i.e.*, heterogeneous contrast enhancement and necrosis), the outgrowing pattern and absence of adjacent tissue invasion made us think of GIST as the primary diagnosis.

The stomach appeared to be distended (Fig. **5**). There was no sign of inflammation or lymphadenopathy. There was no evidence of obstruction preventing the passage of opaque oral material through the stomach and small intestine lumen. Surgical exploration revealed that the mass originated from the second part of the duodenum and fistulized to the duodenal lumen. There was no invasion to the adjacent tissue, and the mass and the anterior wall of the duodenum were excised.

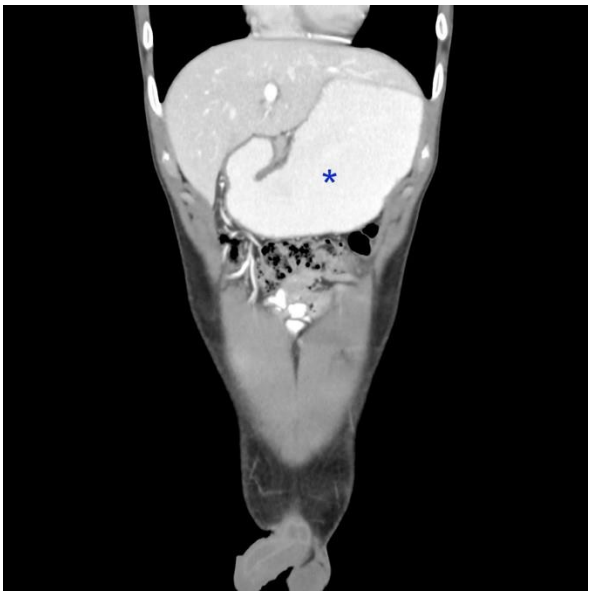

Fig. (5). Distended stomach due to duodenal GIST.

On macroscopic examination, a soft encapsulated mass with bleeding and necrosis areas was observed. On microscopic examination, epithelioid and spindle cells were present. The number of mitoses was 4 at 25 high magnification fields. It had cytological atypia. Tumor cells were stained strongly with CD 117, DOG1, and vimentin. Pathological diagnosis was reported as gastrointestinal stromal tumor (high risk of progressive disease according to Miettinen-Lasota-AFIP data, Prognostic Group 3a). Surgical resection is the standard gold treatment for GISTs. Tyrosine kinase inhibitors are used as adjuvant treatment for patients with high-risk characteristics.It is recommended that patients with high-risk characteristics should be assessed every three months for five years for the possibility of recurrence or metastasis [1]. In our case, as the histopathologic findings suggested a high risk for progression, imatinib treatment was initiated. Abdominal tomography obtained 1.5 months after the surgery demonstrated fat necrosis and some postoperative changes at the surgery site but no residual mass or recurrence, and he was scheduled for regular follow-up.

3. DISCUSSION

GISTs were first defined by Mazur and Clark in 1983 as a subgroup of mesenchymal tumors [6]. However, it was later understood that they arose from the precursors of Cajal Interstitial cells, which are located in the wall of the gastrointestinal tract [7]. It has malignant potential due to oncogene mutation in tyrosine kinase receptor (KIT) and platelet-derived growth factor receptor-alpha (PDGFR-alpha) [8, 9]. CD117 expression is one of the most important immunohistochemical markers that identify GISTs and differentiate them from other mesenchymal neoplasms [10, 11].

According to prognostic factors, 4 risk groups have been defined by the National Institutes of Health consensus classification system: A very low, low, intermediate, and high-risk group. The determination of these groups is based on the long diameter of the tumor, the number of mitoses at 50 magnification, tumor localization, and tumor rupture [12, 13].

Contrast-enhanced CT is the preferred diagnostic method for staging and follow-up. Large tumors have irregular contours, central necrosis, cavitation, and heterogeneous contrast enhancement pattern [14]. Determination of the risk category before treatment is important for the prediction of tumor progression. Several studies show that radiological findings are correlated with the risk category of the tumor. In a study evaluating the CT enhancement pattern and the risk of progression, the heterogeneous enhancement pattern has not been found to be correlated with the histological risk progression of the tumor [15]. Tumors with heterogeneous contrast enhancement patterns are more common in intermediate and high-risk groups, but it has been reported that this may be associated with tumor vascularity, rapid increase in tumor size, and necrosis. Therefore, the heterogeneous enhancement pattern may indirectly reflect the risk of progression [15]. Ulceration is a relatively common finding, especially in duodenal GISTs [3].

Some radiological findings distinguish duodenal and gastric GISTs. In one study, endoluminal growth pattern was seen more frequently in Gastric GISTs [16]. In another study [3], endoluminal growth pattern was not seen in cases of duodenal GIST. It has been proposed that the possible reason for this is that the duodenal lumen is narrower than the gastric cavity, and the mass reaches large dimensions [3]. In the same study, the mean tumor attenuation values in the portal venous phase were found to be higher than gastric GISTs in duodenal GISTs. It has been suggested that this finding may be related to the high blood supply of duodenal GISTs [3]. For this reason, small-bowel-derived GISTs might be more aggressive than gastric-derived GISTs [17, 18].

In differential diagnosis, duodenal adenocarcinoma, pancreatic head adenocarcinoma, pancreatic neuroendocrine tumors, duodenal lymphoma, leiomyoma, paraganglioma, adenoma, and also, aneurysms originating from adjacent arteries in this region and duodenal diverticulum should be considered. In the literature, similar to our report, a case with a diverticulum-like mass accompanied by fistulization to the duodenal lumen has been described [19]. Our case was younger, and the lesion size was very big, with a heterogeneous contrasting pattern suggesting rapid tumor growth. Because of these properties, we aimed to share and discuss this case along with a differential diagnosis and literature review.

CONCLUSION

Duodenal GISTs have a prominent blood supply. It can reach large sizes and can cause necrosis and cavitation. They may show heterogeneous enhancement on CT. Intraluminal gas and ulceration are relatively common in duodenal GISTs. They can be connected with the lumen of the duodenum and form a giant diverticulum. GISTs should be considered in the differential diagnosis of duodenal masses. We presented a young case with a giant duodenal GIST with differential diagnosis and reviewed some diagnostic properties.

LIST OF ABBREVIATIONS

CT	=	Computerized Tomography
GIST	=	Gastrointestinal stromal tumor
KIT	=	Tyrosine kinase receptor
PDGFR-alpha	=	Platelet-derived growth factor receptor-alpha

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Not applicable.

HUMAN AND ANIMAL RIGHTS

No animals/humans were used for studies that are the basis of this research.

CONSENT FOR PUBLICATION

The written informed consent form was taken from the patient.

STANDARDS OF REPORTING

CARE guidelines were followed.

AVAILABILITY OF DATA AND MATERIALS

Not applicable.

FUNDING

None.

CONFLICT OF INTEREST

The author declares no conflicts of interest, financial or otherwise.

ACKNOWLEDGEMENTS

Declared none.

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