OPEN ACCESS

ISSN: 2676-7473

RBMS.2022;27(1):e3

CASE REPORT

Duodenal GIST mass with big management challenges

Mohammad Hassani[1], Mehdi Tavallaei[2], Parynaz Parhizgar[3], Peyman Bakhshaei Shahrbababki[4*]

1. *Assistant Professor of Vascular Surgery, Department of Vascular and Endovascular Surgery, Ayatollah Taleghani Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran. ORCID: 0000-0002-8671-5191*
2. *General Surgeon, Department of General Surgery, Ayatollah Taleghani Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran. ORCID: 0000-0001-7565-1623*
3. *Student Research Committee, School of Medicine, Shahid Beheshti University of Medical Sciences, Tehran, Iran. ORCID: 0000-0003-2651-9786*
4. *Vascular Surgeon, Department of Vascular and Endovascular Surgery, Ayatollah Taleghani Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran. ORCID: 0000-00034936-8685*

*Corresponding Author:
Address: Department of Vascular and Endovascular Surgery, Ayatollah Taleghani Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran.
Email: dr.bakhshaei@gmail.com
ORCID: *0000-00034936-8685*

Date Received: March, 2021 *Date Accepted: April, 2021* *Online Publication: January 15, 2022*

Abstract

Introduction: Gastrointestinal stromal tumor (GIST) is a rare mesenchymal tumor of the gastrointestinal (GI) tract that usually occurs in the stomach and the small intestine, respectively. This tumor often occurs in people over the age of 40 years, especially in the fifth and sixth decades of life. The range of symptoms varies from asymptomatic to symptomatic depending on the size and location of the tumor. The standard diagnostic of this tumor is the pathological examination of tissue specimens. Also today, the main treatment for this tumor is surgery and resection of the tumor.
Case Report: A 43-year-old woman presented with melena and a decrease in hemoglobin level to 4. The patient underwent upper and lower GI endoscopy, CT angiography, and EUS, which was reported a vascular lesion below the ampulla of vater. After laparotomy and Whipple procedure, pathology reported tumor as GIST. The patient was discharged from the hospital in good general condition.
Conclusion: we suggest that patients presented with gastrointestinal bleeding and the presence of a lesion in the diagnostic modalities, GIST should always be considered as a differential diagnosis.

Keywords: Gastrointestinal stromal tumor, GIST, Gastrointestinal bleeding

Introduction

A gastrointestinal stromal tumor (GIST), first introduced in 1983, is known as a rare gastrointestinal mesenchymal tumor (1). The tumor often occurs in the stomach (50%60%) and small intestine (30%-35%). However, this tumor has been reported outside the gastrointestinal tract rarely (2-4). Activation of the mutation in the proto-oncogene c-KIT or platelet-derived growth factor receptor alpha polypeptide (PDGFRA) leads to the formation of this tumor originating in the interstitial cells of the gastrointestinal tract, namely the Cajal, which are responsible for the peristaltic movements of the gastrointestinal tract. This tumor often occurs in people over the age of 40 years, especially in the fifth and sixth decades of life (5). The range of symptoms varies from asymptomatic to symptomatic depending on the size and location of the tumor. In symptomatic cases, patients may experience symptoms such as nausea, vomiting, abdominal pain, gastrointestinal bleeding, and anorexia (6, 7). For a definitive diagnosis, pathology and immunohistochemistry of the tumor are required. Also, the main treatment for this tumor is resection surgery (8).

Case Report

The patient was a 43-year-old woman with melena and a decrease in hemoglobin level to 4 that was referred to Taleghani Hospital in Tehran. After stabilizing the patient's condition, a clinical history was taken from her. The patient mentioned the duration of melena as 4-6 months. The patient was a non-smoker and did not take any drugs. The patient's past medical history was negative. Also, no significant point was found in her family history. On physical examination, her abdomen was soft and she did not have any tenderness or rebound tenderness and guarding. Also, no pathological point was found in the physical examination. The patient initially underwent an upper and lower GI endoscopy, which was normal. The patient then underwent computed tomography (CT) angiography, which reported a vascular lesion near the second part of the duodenum (D2). Because the lesion was vascular, the patient with a suspicion of arteriovenous malformation (AVM) candidate for angiography and angioembolization.In angiography celiac artery had severe stenosis and the blood supply to the liver was mainly provided by anastomosis of the superior mesenteric artery (SMA) with the gastroduodenal artery (GDA), and the vascular lesion also received blood from SMA (Figure 1). Therefore, due to the need to maintain blood supply to the liver and possibility of damage to SMA angioembolization was canceled. The patient underwent endoscopic ultrasonography (EUS) for further evaluation. EUS report showed an external lesion below the major papilla without duodenal mucosal ulcer which pushing inward.

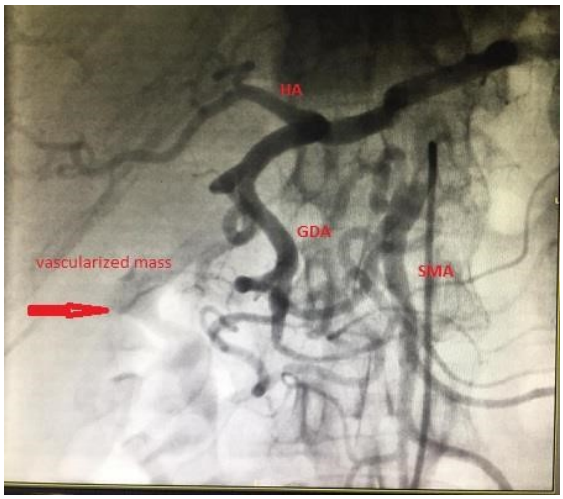

Figure 1: Angiography (HA: hepatic artery, GDA: gastroduodenal artery, SMA: superior mesenteric artery)

The patient was candidate for laparotomy. During surgery, it was found that the major papilla was surrounded by the tumor. Therefore, according to the patient's condition, i.e. stenosis of the celiac artery and the need of the liver for the GDA to supply the blood required for it, the Whipple procedure or pylorus-preserving pancreatoduodenectomy was performed, and also the patient's GDA was preserved. The sample sent to the pathology was a pink to dark red piece with dimensions of 4 cm × 2.8 cm × 2.2 cm, which was reported GIST and had no metastasis (which limit to submucosa and subserosa) . Based on microscopic finding, sections show a biphasic neoplasm compused of uniform eosinophilic cells in short fascicles and whorls, indistinct cell borders, paranuclear cytoplasmic vaculoles and rounded cells with variably eosinophilic cytoplasm in nest and sheet in the

hyaline background. Rare mitotic activity was presented (mitotic rate of 2 per 5mm^2). Tumor histologic grade and pathologic stage was G1 (low grade) and pT2N0Mx, respectively (ICD-O code: C:24.1 M: 8936.1). Figure 2 shows the excised tumor. chronic cholecystitis was reported on pathology report, too. After surgery, the patient's symptoms resolved and the patient was discharged from the hospital in good general condition.

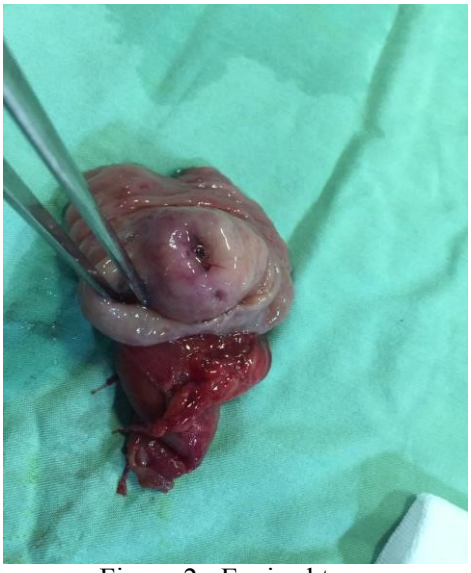

Figure 2: Excised tumor

Discussion

A gastrointestinal stromal tumor (GIST) is known as one of the rare gastrointestinal tumors. The location of this tumor is generally reported in the stomach and then in the small intestine. However, in rare cases, it can be seen outside the gastrointestinal tract (1-4). Gene mutations, especially the proto-oncogene c-KIT or platelet-derived growth factor receptor alpha polypeptide (PDGFRA), and age (because the majority of these tumors are reported to be over 40 years old) have been identified as risk factors for this tumor (5, 9). This tumor is often asymptomatic, although depending on the size and location of the tumor it can be symptomatic, which gastrointestinal bleeding is the most common symptom (6, 7). The patient in this case report also had this symptom.

Diagnostic modalities such as CT scan, MRI, endoscopic ultrasound (EUS), GI endoscopy, and biopsy can be used. However, in cases where the tumor can be resected, the biopsy is not recommended because it carries risks such as bleeding, rupture of the tumor, and metastasis of tumor cells (6). The definitive diagnosis of this tumor is obtained by examination of pathology and immunohistochemistry, usually with the help of CD 117 marker (in 95% of cases of GIST is positive) (10-12). This tumor can metastasize to the liver, omentum, and peritoneal cavity. Lymphatic metastasis is rare (13). Tumor risk for metastasis and rupture is measured based on tumor size and mitotic index. In high-risk cases, it is recommended to treat the tumor with imatinib in addition to surgical treatment. The main treatment for the tumor is still resection of the tumor with surgery. The patient in this case report was placed in a low-risk classification based on the Asian consensus of guidelines (8), so after surgery and examination for tumor metastasis, there was no need for treatment with imatinib.

Indications for the Whipple procedure include a tumor in the head of the pancreas or other structures, including the duodenum, bile duct, or ampulla. In this technique, the head of the pancreas, the duodenum, the first 15 cm of the jejunum, and the gallbladder are removed (12, 14, 15). According to research, if this operation is performed by a skilled surgeon, the mortality rate is less than 4% (16). As mentioned, the patient's celiac artery had stenosis and therefore the blood supply to the liver was provided by anastomosis of the GDA with the branches of the superior mesenteric artery (SMA). Therefore, the Whipple procedure was required to remove the tumor, but the GDA was preserved.

In this case report for the first time in Iran, we reported a 43-year-old patient with GIST and celiac artery stenosis who was successfully treated with surgery. Finally, we suggest that for patients presented with gastrointestinal bleeding and the presence of a lesion in the diagnostic modalities, GIST should always be considered as a differential diagnosis.

Conflict of interest

Authors declare no conflict of interest.

References:

1. Kramer K, Siech M, Sträter J, Aschoff A, Henne-Bruns D. GI hemorrhage with fulminant shock induced by jejunal gastrointestinal stromal tumor (GIST) coincident with duodenal neuroendocrine carcinoma (NET)+ neurofibromatosis (NF)--case report and review of the literature. Zeitschrift fur Gastroenterologie. 2005;43(3):281-8.
2. Choi H. Response evaluation of gastrointestinal stromal tumors. The oncologist. 2008;13:4-7.
3. Joensuu H, Vehtari A, Riihimäki J, Nishida T, Steigen SE, Brabec P, et al. Risk of recurrence of gastrointestinal stromal tumour after surgery: an analysis of pooled population-based cohorts. The lancet oncology. 2012;13(3):265-74.
4. Miettinen M, Lasota J. Gastrointestinal stromal tumors–definition, clinical, histological, immunohistochemical, and molecular genetic features and differential diagnosis. Virchows archiv. 2001;438(1):1-12.
5. Sornmayura P. Gastrointestinal stromal tumors (GISTs): a pathology view point. J Med Assoc Thai. 2009;92(1):124-35.
6. Khourgami Z, Mahmoudzadeh H, Khaheshi I, Bagheri M, Bagheri M, Aminian A. Gastrointestinal Stromal Tumors (Review of Literature). 2008.
7. Tada Y, Yamamoto M, Sawata S, Hara K, Sugesawa K, Ueshima C, et al. Ruptured Small Intestinal Stromal Tumor Causing Concurrent Gastrointestinal and IntraAbdominal Hemorrhage: A Case Report. Yonago acta medica. 2021;64(1):137-42.
8. Nishida T. Asian consensus guidelines for gastrointestinal stromal tumor: what is the same and what is different from global guidelines. Translational gastroenterology and hepatology. 2018;3.
9. Fletcher CD, Berman JJ, Corless C, Gorstein F, Lasota J, Longley BJ, et al. Diagnosis of gastrointestinal stromal tumors: a consensus approach. International journal of surgical pathology. 2002;10(2):81-9.
10. Greenson JK. Gastrointestinal stromal tumors and other mesenchymal lesions of the gut. Modern Pathology. 2003;16(4):366-75.
11. Miettinen M, Sobin LH, Sarlomo-Rikala M. Immunohistochemical spectrum of GISTs at different sites and their differential diagnosis with a reference to CD117 (KIT). Modern pathology. 2000;13(10):1134-42.
12. Tworek JA, Goldblum JR, Weiss SW, Greenson JK, Appelman HD. Stromal tumors of the abdominal colon: a clinicopathologic study of 20 cases. The American journal of surgical pathology. 1999;23(8):937.
13. DeMatteo RP, Lewis JJ, Leung D, Mudan SS, Woodruff JM, Brennan MF. Two hundred gastrointestinal stromal tumors: recurrence patterns and prognostic factors for survival. Annals of surgery. 2000;231(1):51.
14. Duffy JP, Hines OJ, Liu JH, Ko CY, Cortina G, Isacoff WH, et al. Improved survival for adenocarcinoma of the ampulla of Vater: fifty-five consecutive resections. Archives of Surgery. 2003;138(9):941-50.
15. Ryder NM, Ko CY, Hines OJ, Gloor B, Reber HA. Primary duodenal adenocarcinoma: a 40-year experience. Archives of surgery. 2000;135(9):1070-4. 16. Cameron JL, Riall TS, Coleman J, Belcher KA. One thousand consecutive pancreaticoduodenectomies. Annals of surgery. 2006;244(1):10.