Govaresh/ Vol. 27, No.3, Autumn 2022; 194-198.

A Rare Case Report of Gastrointestinal Stromal Tumor Synchronous with Splenic Metastasis

Case Report

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ABSTRACT

A gastrointestinal stromal tumor (GIST) is the most common intramural mesenchymal tumor of the gastrointestinal tract. Their metastasis to extragastrointestinal organs is rare, especially the spleen. The patient was a 52-year-old man who, with the diagnosis of peritonitis, underwent laparotomy emergently. At the time of surgery, an extensive tumor was seen in the stomach, esophagus, and jejunum, which had metastases to the spleen. Total gastrectomy, esophagojejunostomy, and splenectomy were performed. The morphological studies showed a spindle-cell type of GIST that were positive for CD117 and Discovered on GIST-1 (DOG1). GIST was diagnosed as high-risk and treated with imatinib. In follow-up, he died 5 months after discharge. Here, we reported a case of GIST synchronous with splenic metastasis. Surgeons must alert for possible metastases associated with GISTs, clinical presentation, diagnosis, treatment, and pathological classification.

Keywords: Gastrointestinal stromal tumor; GIST; Splenic metastasis

please cite this paper as:
Baradaran R, Mohammadi F, Rahimi Anbarkeh F, Sadeghian A, Sajjadi SH. A Rare Case Report of Gastrointestinal Stromal Tumor Synchronous with Splenic Metastasis. *Govaresh* 2022;27:194-198.

INTRODUCTION

Gastrointestinal stromal tumors (GISTs), which are the most frequent type of mesenchymal tumors, involve the digestive tract. There is a hypothesis that GIST originates from the interstitial cells of Cajal, and hyperplasia of these cells within the muscular wall of the gastrointestinal (GI) tract can induce discrete tumor masses. In addition, the expression of some common genes such as CD117 (KIT), DOG-1, ETV1, PKC-q, and nestin between both ICCs and GISTs confirms this hypothesis (1,2).

GISTs are infrequent compared with other neoplasms present in the GI tract. These tumors can be considered mild lesions with or without metastasis (3-5). The most common source of GIST is the stomach, small intestine, large intestine, rectum, and esophagus, respectively (6-8). There are various signs of GISTs, such as abdominal pain (9), intestinal obstruction (10), peritonitis (11), and hematemesis (12).

In order to differentiate GIST from other tumors in the GI tract, there are a number of specific markers that confirm GIST with high certainty. Examination of CD117, DOG-1, and CD34 markers is essential for the diagnosis of GISTs and their differentiation from other tumors such as leiomyosarcoma. In histological images, GISTs are observed as a monotonous population of spindle cells. Also, in some images, there are observed epithelioid cells or a mixture of epithelioid cells and spindled cells (2,13,14).

Extra GI GISTs are rare. Its most common site is the liver (65%), and the rarest is the spleen (15,16). According to the Updating Consensus Surgical Case Report

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Received: 22 Jun. 2022
Revised: 15 Sep 2022
Accepted: 16 Sep 2022

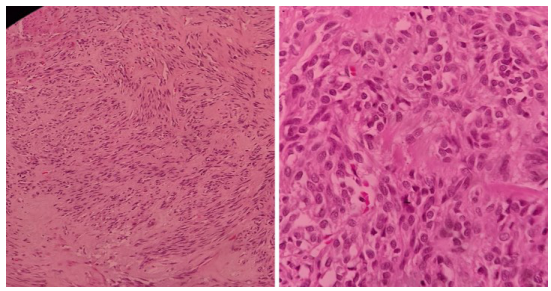

Figure.1: **Histochemistry results showed spindle-shaped cells with oval to elongated nuclei were proliferated and categorized in the fascicles. Also, lymphocytes were infiltrated in the stroma.**

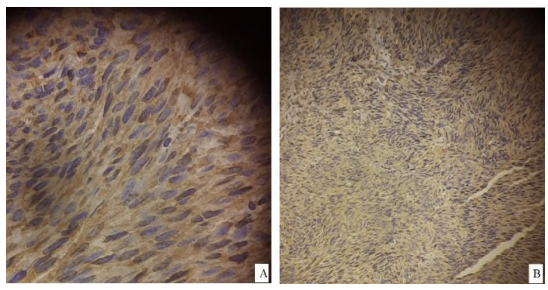

Figure.2: **Immunohistochemistry results showed positive CD117 (A) and DOG1 (B).**

(SCARE) guidelines (17), this case report describes a rare position with GIST within the stomach, jejunum, and esophagus that also metastasized to the spleen.

CASE REPORT

The patient was a 52-year-old man who was referred to our hospital complaining of abdominal pain, fever, chills, shortness of breath, and GI bleeding. The patient's vital signs included Blood Pressure (BP): 130/73, Pulse Rate (RR): 100, Respiratory Rate (RR):12, and Temperature: 37/8. The results of paraclinical tests included Red Blood Cells (RBC):3/57 mm3, Hemoglobin (Hb): 10/4 mg/dL, Hematocrit (Hct): 30/7%, White Blood Cells (WBC): 17600 mm3, C-Reactive Protein (CRP): 3+, Albumin (ALB): 3/22 g/dL, Partial Pressure of Oxygen (PO2): 31/8, Oxygen Saturation (SO2):62/3. The ultrasound results showed 400 cc of free fluid containing clots in the left sub diaphragm area. The patient's initial diagnosis was peritonitis. He underwent immediate laparotomy, which showed extensive gastric cancer that also involved the esophagus and jejunum and invaded the spleen. There were several perforations and lacerations in the involved GI tract. Total gastrectomy, esophagojejunostomy, and splenectomy were performed, and a mass ~ 30 cm long was sent to the laboratory for pathology and final confirmation of the type of cancer. Due to enterocutaneous fistula, the patient underwent conservative treatment. Histochemistry results showed spindle-shaped cells with oval to elongated nuclei were proliferated and categorized in the fascicles within muscularis propria. Also, lymphocytes were infiltrated in the stroma (figure 1). The mitotic index was more than 5/50 high-power field (HPF). Furthermore, immunohistochemistry revealed positive results for CD117 and Discovered on GIST-1 (DOG1) (figure 2).

One day after surgery, the patient was followed up by an upright abdomen X-ray. Upright X-ray of the abdomen and pelvis showed fluid outflow and accumulation at the previous splenectomy site in the left upper quadrant (LUQ) without entering the intestine and leaving the left drainage site in the abdominal wall (figure 3).

GIST was diagnosed as high-risk and treated with imatinib mesylate (400 mg daily). The patient was discharged, treatment was continued, and he was followed up. Unfortunately, he died 5 months after discharge.

DISCUSSION

In the present study, we report a case of GIST

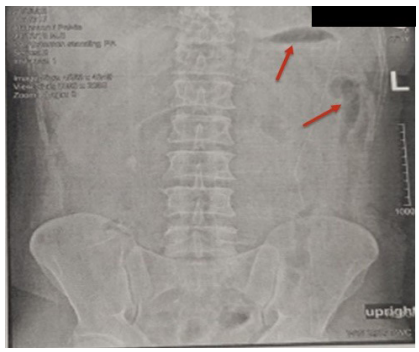

Figure.3: one day after surgery, an upright abdominal radiograph shows fluid accumulation at the previous splenectomy site (air-fluid levels sign) (arrows) and the LUQ region without entering the intestine and leaving the left drainage site in the abdominal wall.

Table 1: Summary of Health Information Technology application in Celiac Disease Diagnosis

Authors	Year	Age	Sex	Diagnostic method	Treatment	Pathological diagnosis	Mitotic index	Tumor size
Li et al.	2011	56	F	CT scan, physical examination	Local excision of gastric lesions, splenectomy	CD117, CA125	<5/50 HPF	3.5 cm
Kim et al.	2011	59	M	CT scan, physical examination	Previous gastric wedge resection, splenectomy	CD117, CD34	>10/50 HPF	12.2 × 11 cm
Palanivelu et al.	2016	67	F	Endoscopy, CT scan, physical examination	Segmental gastric resection, splenectomy, excision of diaphragm cuff	CD117, CD34	6/50 HPF	>10 cm
Baradaran et al.	2022	52	M	-	Total gastrectomy, esophagojejunostomy, and splenectomy	CD117, DOG1	>5/50 HPF	~ 30 cm

in the stomach, jejunum, and esophagus that had metastasized to the spleen. In our best search of the literature, there are few reports of GISTs metastasis to the spleen.

GISTs are mostly benign and less likely to metastasize to other organs. Because of their rarity, published literature mostly are case reports. GISTs occur in approximately 10-20/ per one million people/ per year. They can be seen anywhere from the GI and the intra-abdominal organs. But the most common place is the stomach (70%) and small intestine (10-20%); also, concomitant tumors are rare (18). Furthermore, among the intra-abdominal organs, the most common metastatic site is the liver (65%) (15), and the rarest is the spleen (16).

In our best search in English literature, there are only three cases that have reported GISTs, which had metastasized to the spleen (presented in table 1). Li and colleagues reported a rare case of GIST in a 56-year-old woman that was metastasized to the spleen and synchronized with ovarian carcinoma. Splenic metastases were placed on the splenic capsule and were spread through the peritoneum. Finally, he underwent resection of gastric lesions and splenectomy (19).

Kim and co-workers presented a case of recurrent gastric GIST that was metastasized to the spleen 14 months after removing GIST of the gastric cardia. They attributed the treatment failure and tumor recurrence to the capsular tearing and tumor seeding during the first operation (16).

Palanivelu and others explained a scarce case of a malignant gastric GIST which involved the spleen and diaphragm. In the endoscopic view, they observed a small tumor because most of the tumor had grown exophytically, and had stuck to the diaphragm in the upper direction and to the spleen in the lateral direction. Finally, splenectomy and shaving of the diaphragm tissue were performed until a healthy margin was reached (20).

The synchronous of gastric GIST and other organ tumors may be related to environmental agents such

as helicobacter pylori infection, carcinogenic regimen or abuse of drugs that oppose epithelial and stromal tissues, specific genetic aberration, tumor growth among a specific population, or/and iatrogenic agents (21,22).

Symptoms of the present patient included abdominal pain, fever, chills, shortness of breath, and GI bleeding. It has been reported that GISTs are asymptomatic in the early stages because they grow toward the lamina propria of the muscle and appear as masses outside the organs and do not often cause bleeding or obstruction inside the lumen (23). However, in more advanced stages, signs and symptoms of GIST are abdominal pain, palpable mass, GI bleeding, fever, anorexia, weight loss, and anemia. GISTs may perforate and induce peritonitis, which is rare. Intestinal obstruction may also occur (24,25).

The most important prognostic indicators for GISTs are mitotic index and tumor size (26), which can divide tumors into four classes: very low risk, low risk, intermediate risk, and high risk (27). Based on the clinical, surgical, and histopathological observations, the present patient was placed in the high-risk class for follow-up and the possibility of recurrence.

Histopathologically, GISTs include 70% spindle cell, epithelioid, and round cell or their combination, which are located in extremely compressed fascicular with pale and eosinophilic intracytoplasmic vacuoles and skeinoid fibers (28). The final and definitive confirmation of the GIST is the immunohistochemical tests for CD117 antigen and DOG1 (29,30). This was done for our patient, and the results showed positive CD117 and DOG1.

The best treatment for patients with GIST is complete surgical resection until a clear margin is reached. Imatinib has also been suggested as adjunctive therapy after GIST (31,32). In the present patient, these treatments were also performed on the patient, and he was followed up, but unfortunately, he died after 5 months.

In conclusion, GISTs metastasis to the spleen is rare. Surgeons, radiologists, and pathologists are suggested to consider possible metastases that are associated with GISTs, clinical presentation, diagnosis, treatment, and pathological classification.

CONFLICT OF INTEREST

The authors declare no conflict of interests related to this work.

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