A case of gastrointestinal bleeding secondary to a small bowel gastrointestinal stromal tumor before causing hemoperitoneum

Gastrointestinal stromal tumors (GISTs) is a rare stromal neoplasm that represent the most common mesenchymal tumor of the GI tract, accounting for 0.2% of all GI malignancies and 5% of all sarcomas.\(^1\) GIST originating from interstitial cells of Cajal, which are regulators of gut peristalsis, are preferentially located in the stomach and the small intestine.\(^2\) It is characterized by indolent clinical symptoms including vague abdominal pain, weight loss, GI bleeding and obstruction, which is caused by the growing tumor. The usual presentation of GIST is a GI bleed. Acute presentation of GIST with features of hemoperitoneum has never been reported. We report an unusual case of small bowel GIST in which the patient presented with GI bleed before presenting as hemoperitoneum.

**Key words:** Exophytic, gastrointestinal stromal tumor, hemoperitoneum, small bowel

**INTRODUCTION**

Gastrointestinal stromal tumors (GISTs) is a rare stromal neoplasm that represent the most common mesenchymal tumor of the GI tract, accounting for 0.2% of all GI malignancies and 5% of all sarcomas.\(^1\)\(^-\)\(^2\) About 70% occur in the stomach, 20% in the small intestine and less than 10% in the esophagus. They rarely occur in other abdominal organs. GISTs are thought to arise from interstitial cells of Cajal,\(^3\) that are normally part of the autonomic nervous system of the intestine. They serve a pacemaker function in controlling motility.

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**CASE REPORT**

A 45-year-old man presented to our emergency department with a week-long history of dull aching lower-abdominal pain and no other symptoms. The severity of pain had increased over the last 2 days and was accompanied by progressive abdominal distension and vomiting. On clinical examination, the patient was pale and diaphoretic and had tachycardia with systolic blood pressure 90 mmHg. Abdominal examination showed abdominal distension, diffuse abdominal tenderness, guarding, and rigidity. Routine laboratory investigation revealed Hb-6 gm%. Abdominal X-ray (erect) not showed gas under right dome of diaphragm. Ultrasound examination showed a 5 × 6 cm mass originating from small bowel with free fluid in the abdomen noted. USG-guided free fluid aspiration shows blood after repeated aspiration. The patient was hemodynamically stabilized and conservatively managed, and later planned for contrast-computed tomography of the abdomen for detailed evaluation.

Apart from the above he was a healthy male presented with hematochezia and melena since 6 months on and off without any co-morbidity and no history of previous abdominal surgery. Initial investigations included an abdominal ultrasound scan (USG), an esophago-gastro-duodenoscopy (OGD) and flexible
sigmoidoscopy and colonoscopy were also unremarkable. His malena and hematochezia thought due to healed gastric or duodenal lesion and he was advised to take proton pump inhibitors on a regular basis, which resulted in partial and temporary relief of his symptoms. Although his condition did not impact his lifestyle significantly, his symptoms never subsided completely and almost 2 months following initial presentation repeat OGD and colonoscopy not revealed anything. All of these did not reveal any abnormality and thus planned for diagnostic laparoscopy, but the patient was hesitant and refused for surgery. Four months later the patient presented with features of hemoperitoneum.

Finally, 4 months from presentation, the patient was admitted to hospital with hemoperitoneum and underwent a laparotomy within 48 hours of admission after hemodynamic stabilization with adequate fluid and electrolyte replacement. Blood transfusion was done to make case fit for surgery. A clinical diagnosis of hemoperitoneum probably due to gastroduodenal lesion or rupture of intraabdominal abscess was made, and emergency laparotomy was planned.

On exploration of the abdomen, hemoperitoneum noted of around 500 ml and 5 × 6 cm exophytic mass [Figure 1] originating from the proximal jejunum about 30 cm distal to the duodenojejunal flexure. It was a firm, lobulated mass with areas of softening and cystic degeneration with overlying distended veins and small intratumoral hematoma [Figure 2]. The tumor was hypervascular, with evidence of recent hemorrhage into the tumor. No evidence of visceral metastasis was found. En bloc excision of the tumor with jejunojejunal anastomosis was performed. Postoperative recovery was uneventful.

Histopathological analysis revealed a densely crowded spindle and polygonal cell tumor showing moderate nuclear pleomorphism [Figures 3]. 3-4 mitoses/HPF were found and the tumor spread into the mucosa and outwards into the serosa. There was no lymph nodal spread. Immunohistochemistry demonstrated positive staining for CD117 (cKit) and negative staining for CD34, S-100 and chromogranin. Based on the size (5 × 6 cm) and mitosis (3-4/HPF), the tumor was classified as a gastrointestinal stromal tumor (GIST) of intermediate malignant potential. A CT scan of the chest, abdomen and pelvis performed subsequently did not reveal the presence of metastatic disease. The patient was discharged on the 10th postoperative day and decided not to proceed with any adjuvant treatment after departmental meeting. The patient made an uneventful recovery and was remarkably well after 1 year of follow up; all his previous abdominal symptoms were completely resolved.

**DISCUSSION**

GISTs are rare neoplasms of the GI tract ranking a distant third after adenocarcinomas and lymphomas. GISTs can occur anywhere in the GI tract. Although morphologically similar to other benign and malignant smooth muscle and neural stromal tumors, GIST constitutes a distinct group of rare GI tract tumors that originate from the interstitial cells of Cajal. The latter are regulators of gut peristalsis and normally express CD117, which is a product of the c-kit proto-oncogene that encodes a tyrosine kinase receptor, which regulates cellular proliferation in GIST.
GISTs arise from the muscularis mucosa or muscularis propria layers and most exhibit an endophytic growth pattern, growing within the bowel lumen. The overlying mucosa is usually preserved but larger and more aggressive tumors tend to ulcerate through this. They are submucosal lesions that often grow intraluminally. Sometimes they have an extraluminal exophytic component. The stomach (60%) is the most common site followed by small intestine (30%), duodenum (5%), colon/rectum (5%) and esophagus (<1%). Primary mesenteric, omental and retroperitoneal GISTs have also been reported, but they are quite rare.[5]

GIST is observed predominantly in adults at a median age of 58 years. The incidence is equal in men and women. It rarely presents in younger age, but in our case, the patient presented at the age of 45 years. Clinical presentation of patients varies according to site, size and aggressiveness of the tumor. Symptoms manifest when the tumor is large (>5 cm) or situated in a critical anatomical location (e.g. causing gastric outlet obstruction). Symptoms may include abdominal pain, mass, nausea, vomiting, anorexia and weight loss. Patients may present with GI bleeding (due to pressure necrosis and ulceration of the overlying mucosa) or with obstructive symptoms. An extensive review of the literature regarding GISTs not revealed any tumor presenting as hemoperitoneum. Two GISTs out of which one was a gastric perforation and the second one was jejunal[6,7] but no tumor presented as hemoperitoneum. So, ours is the only reported case presenting as jejunal tumor presenting as exophytic growth with hemoperitoneum. Definitive surgery remains the mainstay of treatment for patients with localized primary GIST.

CONCLUSION

Due to the relative inaccessibility of the small bowel, diagnosis of small intestinal GI bleeding remains a challenge. Advances in imaging techniques of the small bowel allow us to better evaluate areas that have previously been poorly visualized. Video capsule endoscopy have a role in the evaluation of patients presenting with GI bleeding and should be considered as a diagnostic option in the assessment of such patients, especially in those patients in whom no source can be clearly identified. Clinicians should consider it to use as a complementary diagnostic option to OGD and colonoscopy in the evaluation of small intestinal GI bleeding although a diagnostic laparoscopy and CT scan may have helped detect the tumor when very small which was initially missed by USG. It could have been considered at some stage during the months preceding his final presentation as hemoperitoneum.

REFERENCES