Hemorrhage, Intestinal Angina & Ectopic Pregnancy: Atypical Presentations Of Gastrointestinal Stromal Tumors

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Citation

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Abstract

Gastrointestinal Stromal Tumors (GISTs)n are the most common mesenchymal tumors of gastrointestinal tract. Most of the GISTs are asymptomatic. Symptomatic patients usually present with abdominal pain. We describe our recent experience with three patients who presented with unusual symptoms of GIST namely, massive gastrointestinal hemorrhage, intestinal angina and ectopic pregnancy. Stomach is the most common site of occurrence. Discovery of antigen CD 117 has been a major breakthrough in diagnosis of GIST. Prognostic indicators include size of the tumor and mitotic rate. CT scan is the diagnostic study of choice. Surgical resection remains the main treatment modality. Several drug therapies including imatinib are under investigation.

INTRODUCTION

Stromal or mesenchymal neoplasms affecting the Gastrointestinal (GI) tract are divided into two groups. The less common variety includes tumors that are identical to those arise in the soft issues throughout the rest of the body, including lipomas, schwannomas, hemangioms, usual leiomyomas and their malignant counterparts (e.g. leiomyosarcoma) The second more common group consists of stromal tumors that are collectively referred to as gastrointestinal stromal tumors (GISTs). They are most often located in the stomach and proximal small intestine, but can occur in any portion of the alimentary tract that contains smooth muscle within its wall, and occasionally in the omentum, mesentery, and peritoneum[1, 2]. This text will detail our recent experience with three cases of this disease entity and a literature review.

CASE REPORTS

CASE 1

A 45-year-old female presented with complaints of dizziness. She had history of aspirin intake for chronic headaches, and also had tarry stools. She denied abdominal pain, weight loss, nausea or vomiting. On physical examination, her vital signs were stable. Abdominal examination was normal. Hemoglobin on admission was 4.5 gram/ dl with microcytic anemia. Coagulation profile was normal. Patient had 4 units of blood transfusions, and then underwent an endoscopy which showed a 3cm mass in the greater curvature of the stomach with mucosal ulceration. Biopsy was negative for malignancy. Colonoscopy was normal. The patient was started on protonix. CT of abdomen showed 2.6 x 2.3 cm intraluminal filling defect in the distal stomach.

An exploratory laparotomy was done. A 5cm mass was found in the greater curvature of stomach, about 3 cm from the incisura angularis. Gastrocolic omentum was opened and the lesser curvature of stomach was accessed. Distal one third of stomach was devascularized. Following that, just beyond the pyloric point, the duodenum was transected using GIA-75. the stomach at the junction of upper two thirds and distal one third was also transected using the GIA 75. Frozen section of the specimen showed gastrointestinal tumor with no evidence of malignancy. A gastrojejunostomy was then performed. Abdomen was closed. The post operative course was not complicated. The patient was started on clear liquid diet on second post operative day. Diet was gradually advanced, and the patient was discharged home on post operative day 5. Final pathology results showed benign proliferation of smooth muscle cells. These cells were epithelioid in nature round to oval cells and distinct nuclear borders. Nuclei were uniform and benign. There was no evidence of increased mitotic activity. CD117 staining was positive, Reticulin stain showed increase reticulin fiber production. These findings are consistent with gastrointestinal tumor (epithelioid leiomyoma) with overlying mucosal ulceration.

CASE 2

A 67-year-old man presented with complaints of periumbilical pain for 6 months. Pain was associated with food intake. Patient started to avoid food in order to get pain relief, and reported a loss of 30 pounds body weight in 6 month duration. The patient denied any weight loss, fever, anorexia, chronic fatigue, blood in stool, or change in bowel habits. On physical examination, the patient appeared healthy. The abdomen was not distended and was nontender and soft. No ascites, hepatomegaly, or splenomegaly was noted. Laboratory studies revealed normal levels of hemoglobin, hematocrit, bilirbuin, aspartate aminotransferase, and alanine aminotransferase.

An abdominal computed tomography (CT) scan demonstrated a tumor in the mesentery of the small bowel in close proximity to the superior mesenteric artery and vein.

A midline laparotomy was performed and a 6 cm solid mass found in the base of the small bowel mesentery. The lesion was in close proximity to the superior mesenteric artery and vein. There was no evidence of invasion of vascular structures. There were no abnormalities found in the liver, spleen, and pancreas and no lymphadenopathy in the small or large bowel mesentery. Branches of the superior mesenteric artery and vein going to the tumor were occluded with vascular clamps, and the small and large bowel were observed for viability. A 40 cm long portion of the small bowel, in close proximity to the tumor became dusky. The small bowel mesentery tumor was removed by transecting and ligating the previously clamped branches of the superior mesenteric artery and vein. The continuity of the small bowel was restored by anastomosing the transected ends within a GIA-75TM stapling devised in side-to-side fashion, creating and end-to-end functional anastomosis.

The postoperative course was uneventful. The diet was advanced gradually, and the patient resumed having normal bowel movements. He was discharged on postoperative day seven.

Hematoxylin and eosin staining revealed a dense proliferation of spindle cells and inflammation, resembling granulation tissue. Immunohistochemical staining for CD117 was positive. The lesion demonstrated focal cytokeratin staining positively, as well as positive staining for the smooth muscle actin. These features are diagnostic of Gastro intestinal stromal tumor.

CASE 3

A 20-year-old woman who stated that she was 4 weeks pregnant presented with right lower quadrant pain of 2 days duration. The pain was described as dull, increasing with activity but not radiating. She had no other complaints. Her physical examination was normal except for slight tenderness in right lower quadrant. Her CBC, SMA 12, Liver function tests, and urine analysis were normal. Her B-HCG (Humon Chorionic Gonadotropin) level corresponded to the gestational age of the embryo.

Abdominal ultrosonography revealed no intrauterine prengnacy, but showed a 5.7cm, sac-like structure in the right adenexa that was suspicious for ectopic pregnancy. A diagnostic laparoscopy was performed. There were normal fallopian tubes and ovaries bilaterally, but a solid smooth, pedunculated, and greenish mass, measuring 3.2x2.5x2.5cm was identified on the antimesenteric border of the ileum. No other abnormalities were found in the liver, spleen, and pancreas, and there was no lymphadenopathy. About 7cm of the ileum was transected, including the mass in the mesentry, and a primary end-to-end anastomosis was performed 20 cm from the ileocecal valve. The ileocecal valve was preserved. Hematoxylin and eosin staining revealed a dense proliferation of spindle cells and inflammation which resembled granulation tissue. Immunohistochemical staining was positive for cytokeratin, desmin, P53,
S100, CD34, CD117, vimentin, and smooth muscle actin. These features were diagnostic of gastrointestinal stromal tumor.

The patient's postoperative recovery was uneventful. Her diet was advanced as tolerated until she fully regained bowel function. She was discharged to home on postoperative day 5.

DISCUSSION

Mesenchymal tumors are the most common non-epithelial benign tumors of gastrointestinal tract, however they constitute only 1 percent of primary GI cancers[2]. Most of the GIST tumors are asymptomatic. Symptomatic patients present with occult gastrointestinal bleeding (40%), abdominal mass (40%) and abdominal pain (20). [3]CT of abdomen is the diagnostic modality of choice. The usual CT appearance of is that of a solid mass that enhances brightly with intravenous contrast.

Stomach is the most frequent site for GISTs. Small intestine

is the second most frequent site for smooth muscle tumors. Tumors are most commonly found in the jejunum followed by the ileum and the duodenum. For symptomatic patients, surgical resection of the entire tumor is the treatment of choice. Expression of CD117 is diagnostic of GIST.

The clinical behavior of GISTs is variable. Approximately 50 percent of completely resected GISTs tend to recur within the first five years of follow-up[1, 4]. With long term follow up, any GIST presenting with clinical symptoms or signs leading to treatment has the potential to behave in a malignant fashion [5]. Demetri [6] describe that aggressive behavior of GIST depends on tumor size and mitotic count. Size greater than 10cm or mitotic count greater than 10 per 50 high power field dictates the highest risk for clinically aggressive GIST.

The benefit of adjuvant imatinib is still under clinical investigation. Duration of treatment and follow up after therapy is not clear at this point. For patients with nonmetastatic unresectable GISTs, we suggest therapy with imatinib followed by attempted resection, depending on the clinical circumstances. We recommend annual CT of abdomen as a follow up after surgical resection.

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