A rare presentation of a Leiomyosarcoma of the Stomach

P Saxena, M Nanda

Citation

P Saxena, M Nanda. *A rare presentation of a Leiomyosarcoma of the Stomach*. The Internet Journal of Surgery. 2006 Volume 9 Number 2.

Abstract

Gastrointestinal stromal tumors {GIST} are very rare tumors of GIT [0.5-3% of all stomach malignancies] evenly distributed between stomach and small intestine. They commonly present with bleeding, abdominal pain, abdominal fullness and discomfort, anemia, palpable abdominal mass, ulceration etc. Tumor perforation is very rare. Here we are reporting a case, which presented in emergency with gas under diaphragm. On exploration a very huge tumor mass arising from stomach with areas of necrosis and gas bubbles was found. There was no perforation of any hollow viscus. Histopathology revealed epitheliod leiomyosarcoma with degeneration cysts.

CASE PRESENTATION

A 60 year old male presented in emergency at surgery department, Gandhi Medical College, Bhopal with complains of pain in abdomen of 2 months duration, low grade fever 2 months, weight loss 1 month, distention of abdomen 15 days and not passing flatus and motions 2 days. Pain was initially in the epigastrium and later on felt in whole of the abdomen. It was dull aching in nature, aggravated with meals but was not associated with vomiting initially. He also had low-grade fever off and on in the last 2 months He had lost weight significantly. About 15 days back he started complaining of distention of abdomen, which was felt after taking meals and was sometimes followed by 1 or 2 episodes of vomiting particularly at night. Vomitus contained food particles, was non-bilious and was nonprojectile in nature. He didn't pass flatus and motions in the last 2 days and distention of abdomen increased further. There was no history of hematemesis, melena or anorexia. Patient was a known asthmatic and has been on medication. He also had chronic constipation and was a tobacco chewer. On admission pulse was 108/min, blood pressure was 120/70 mm Hg and respiration rate was 20/min. The patient was emaciated and looked listless and lethargic. Abdomen was distended and felt rigid and tender all over. Liver dullness was not masked. Bowel sounds were sluggish.

On investigating Hemoglobin was 7 gm%,,Bl. Urea-49 mg%, Serum creatinine was 2.2 mg% X ray abdomen showed gas under diaphragm. USG of abdomen showed free fluid in hepatorenal pouch and feacal filled bowel loops with sluggish peristalsis. A provisional diagnosis of perforation-

peritonitis was made. Despite adequate fluid resuscitation and blood transfusion there was no urine output and the blood pressure shot up to 200/110. So on explained risk an exploratory laparotomy was planned.

On exploration a huge tumor mass was seen extending from epigastrium to pelvis vertically and from left to the right paracolic gutter transversely. It showed areas of necrosis and hemorrhages. Gas bubbles were seen coming from the tumor. The transverse colon was pushed in the pelvis and small intestine was lying below it in the pelvic cavity. Superiorly the tumor was adherent to the inferior surface of liver but there was no infiltration or secondary deposits in the liver. Likewise laterally on the left side it was adherent to the medial aspect of spleen. The tumor was freed from its attachments and was delivered out. Greater omentum was normal and the tumor was found to be attached to the anterior wall of the fundus of the stomach. There was no evidence of perforation of stomach or any other hollow viscus. Taking an adequate margin the fundus of stomach was excised along with the tumor and the defect closed in two layers. After peritoneal toilet the abdominal wound was closed keeping a drain in the pelvis and the Morrisons pouch.

Postoperatively urine output gradually returned to normal. The patient was listless for about 4 days but normalized gradually on correction of fluid and electrolyte imbalance. Oral intake was resumed after 7 days, when the drains were removed. He had superficial wound infection that was controlled. He was discharged from hospital after 3 weeks. Histopathology revealed epitheliod leiomyosarcoma with many degeneration cysts. Tumor size was 25cm x 24cm. Pathology section showed highly pleomorphic cells with clear cytoplasm.

Gastrointestinal stromal tumors {GIST} are considered to be a subset of GI mesenchymal tumors of varying differentiation previously classified as leiomyoma, leiomyosarcoma, leiomyoblastoma, schwannoma etc. There is a consensus that GIST originate from a common primitive cell with dual characteristic of muscle and neuron cells similar to interstitial cell of cajal. These tumors are positive for CD-117 kit protein on immunohistochemical staining. C kit protein is a membrane receptor with a tyrosine kinase component and mutation in CD-117 gene has been linked to malignant potential. Leiomyoma and leiomyosarcomas are myogenic in origin and react positively for smooth muscle actin and negatively for c-kit. Other markers for mesenchymal tumors are CD-34, SMA, 3S100, actin and desmin.

These tumors usually present in fifth to seventh decade, are equally distributed between both sexes, and vary in size from less than 2 cm to 30 cm. Grossly the tumors are solid or cystic with areas of hemorrhages and necrosis, including mucosal ulceration and cavitations. There is a wide range of morphologic expression with spindle, epitheliod and pleomorphic forms. Two principle cell types are spindle and epitheliod forms which may coexist in varying proportions. Larger GIST outgrows its vascular supply leading to extensive areas of necrosis and hemorrhage. The commonest clinical presentation is with bleeding. Other clinical manifestations are abdominal pain, abdominal fullness and discomfort, anemia and palpable abdominal mass (2, 4, 5). The complications are tumor necrosis, ulceration, hemorrhage and tumor perforation. Metastasis is primarily heamatogenic to liver. Lymphatic spread is rare (<< 10%). CT scan with iv and oral contrast is ideal for delineating

both endoluminal and exophytic extent of tumor. GISTS appear as heterogeneous masses with enhancing borders. Irregular central areas of fluid air or oral contrast attenuation may be seen. Treatment is primarily surgical excision with adequate margins. Five years survival has been reported to be 68 to 90% with curative resections. Tumor size, mitotic count and resectability correlated well with tumor recurrence and survival (1). Prognosis is poor in tumors >>10 cm, mitotic count >>10/50 hpf, or with incomplete resection. Poor prognosis has also been reported with tumor rupture, distal location of tumor, high cellularity, and tumor necrosis, presence of metastasis or mutation in C kit gene. Recurrence occurs at primary site, peritoneum or liver. Median survival is 60 months with primary disease, 19 months with metastasis and 10 months with local recurrence. Drugs interacting with tyrosine kinase enzymes like STI-571, Gleevec and Imatinib have been shown to be useful (1, 3). An oncological reference was sought and the patient was advised to come for follow up. Since the disease was mainly exophytic he has to be followed up by repeat USG and CT scans with or without gastroscopy.

ACKNOWLEDGEMENT

Dr Rini Malik , MD Path , Department of Pathology , Gandhi Medical College , Bhopal

References

 Lin SC, Huang MJ, Zeng CY, Wang TI, Liu ZL, Shiay RK. Clinical manifestations and prognostic factors in patients with gastrointestinal stromal tumors . World J Gastroenterol. 2003;9: 2809-2812.
Intestinal leiomyosarcoma, Martinage GE, E-medicine. http://www.emedicine.com/med/topic1180.htm. 29.12.2004
Pathology of Gastrointestinal stromal tumors, Cyril Fisher MD, D.Sc FRCPath , Royal Marsden Hospital ,London.www.pathsoc.org.uk/pathcola/july2003
E-medicine, Gastrointestinal, Dec 11, 2002. Vu Nguyen MD, Andrew Tailor ,MD. University of Wisconsin Hosp, USA
Hemoperitoneum secondary to exogastric Leiomychlostoma of atemach Pasal Al Surnait MD. Leo

Leiomyoblastoma of stomach, Basel Al Surnait, MD, Joe levi, MD. University of Miami, Florida, USA. Annals of surgery, vol 19, No 31/999

Author Information

Pradeep Saxena, M.S [Surgery] Assistant Professor, Department of Surgery, Gandhi Medical College

Monica Nanda, RSO

Department of Surgery, Gandhi Medical College