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Case Report

Malignant Gastrointestinal Stromal tumor of the Sigmoid Colon with Perforation and Peritonitis - an Unusual Presentation

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ABSTRACT

Gastrointestinal Stromal Tumors (GISTs) are the most common subset of gastrointestinal (GI) mesenchymal tumors of varying differentiation. GISTs are defined as mesenchymal, spindle-shaped tumors which can be distinguished from other soft tissue tumors like leiomyomas, myoblastomas etc by C- Kit proto-oncogen (CD - 117) expression. With 60-70%, the stomach is the most common site for a GIST, followed by the small intestine (20-30 %). Colorectal GISTs are frequently rare, accounting for only 5 %. GIST is usually associated with abdominal pain, palpable mass or GI bleeding. However GIST originating from the sigmoid colon rarely causes perforation. Spontaneous rupture is a very rare manifestation of a GIST.

Herein we present a case of a 70 year old male of malignant GIST of the sigmoid colon with perforation which is an extremely rare phenomenon. To the best of our knowledge, this is the second documented case of malignant GIST of colon with perforation and peritonitis.

Keywords: Gastrointestinal stromal tumor (GIST), malignant perforation, sigmoid colon, gastrointestinal tract (GIT).

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are uncommon tumors accounting for 0.1 – 3 % of all gastrointestinal neoplasms, most commonly occurring in stomach (60-70%) or small intestine (20-30 %). [1, 2] Colonic GIST is frequently rare and accounting for only 5 % of tumors arising in the digestive tract. [2] But GIST represents 80 % of gastrointestinal mesenchymal tumors. Moreover the pathobiological features of malignant GISTs of the colon remain unclear. [3] These tumors originate in the interstitial cells of Cajal (GI pacemaker

cells) or more primitive stem cells from which both Cajal cells and smooth muscle cells arise. [4]

Small GISTs are usually asymptomatic, but as the tumor grows- vague symptoms develop such as pain, abdominal discomfort, bloating, mass, and overt or occult bleeding. [4] The most common presentation is gastrointestinal bleeding (acute/chronic) related to mucosal erosion in approximately 50 % cases. [5] Colonic GISTs were typically transmural tumors with frequent intraluminal or outward bulging component. [6]

An extensive review of the literature and pubmed search regarding GISTs revealed only one report of perforation of colonic GIST in 2011 by Hwango Y et al. [4] Herewith we present a second case report as malignant GIST of sigmoid colon with extremely rare presentation as perforation and peritonitis.

CASE REPORT

A 70 year old male presented to the surgical OPD of our hospital with chief complaints of severe pain in abdomen and distention since 2 days. There was no history of fever, vomiting, hematemesis / malena. History of pain in the abdomen was on and off since 6 months. There was no significant personal / family history. Local examination revealed tenderness all over the abdomen with guarding and rigidity. Sluggish peristalsis was noted. Clinical findings were suggestive of intestinal obstruction and the patient underwent emergency radiological examinations. X ray abdomen showed free air under the diaphragm and soft tissue shadow in the pelvic region. USG abdomen showed soft tissue mass in the pelvic region with collection of fluid and sluggish peristalsis most likely arising from sigmoid colon with multiple hypo-echoic shadows suggestive of hollow viscous perforation. In view of these findings, emergency laparotomy was carried out which revealed perforation of sigmoid colon with gray white large mass protruding out through the defect (figure1). Large collection of peritoneal fluid admixed with fecal matter was noted. Wide excision of the sigmoid colon with growth was carried out with sigmoid end colostomy. The resected specimen was sent for histopathological examination. Post operative was uneventful and the patient is doing well on regular follow up.

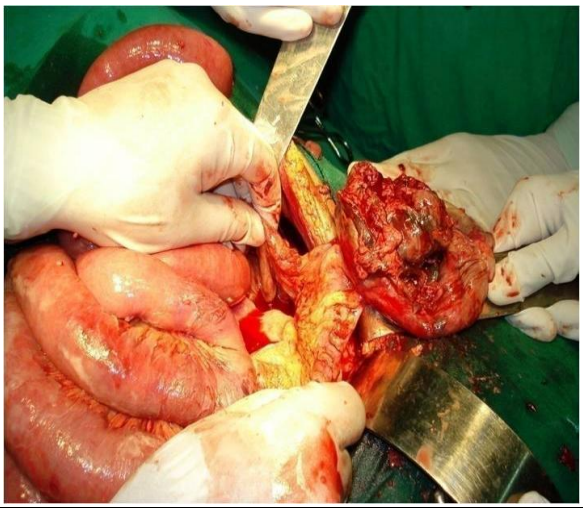

Figure 1: Gross photograph of perforated sigmoid colon with gray white mass protruding out through defect.

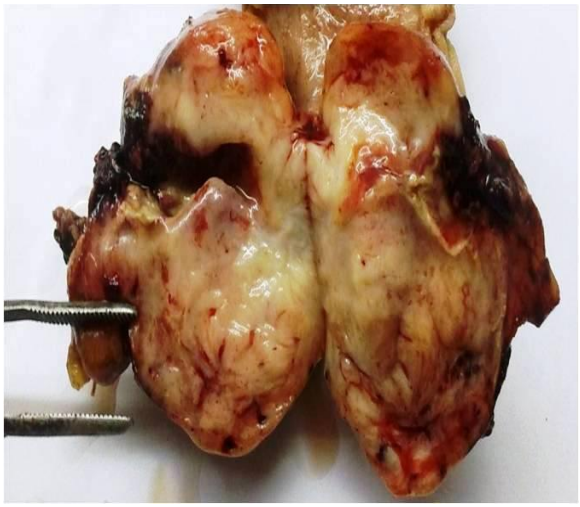

Figure 2: Gross photograph of cut section of tumor mass showing grayish white fleshy appearance with destruction of colonic wall.

Gross findings:

We received left sided ruptured sigmoid colon with grey white mass totally measuring 15 x 10 x 5 cm. Tumor mass was seen in the wall of the sigmoid colon measuring 6 x 6 cm, which was irregular, friable, dark brown with extensive areas of destruction of the wall of the colon. Area of irregular perforation measuring 4 x 3 cm was seen filled with hemorrhagic debris. Cut section of the tumor mass showed grayish white, fleshy appearance with destruction of colonic wall (figure 2).

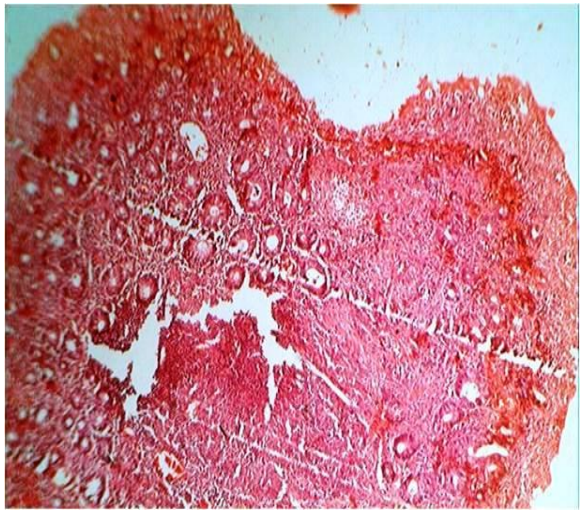

Figure3: Photomicrograph of spindle cell tumor arising from muscularis propria and reaching upto mucosa of colon leading to ulceration.(H & E, x100).

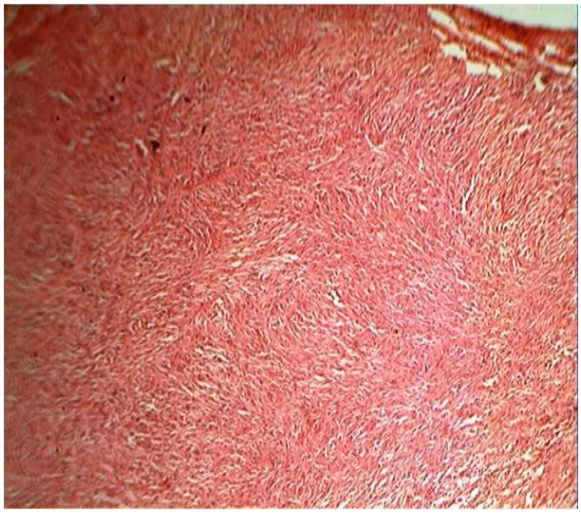

Figure 4: Photomicrograph of tumor showing plump spindle shaped tumor cells arranged in short fascicles, whorls, sheets and nests .(H & E, x100).

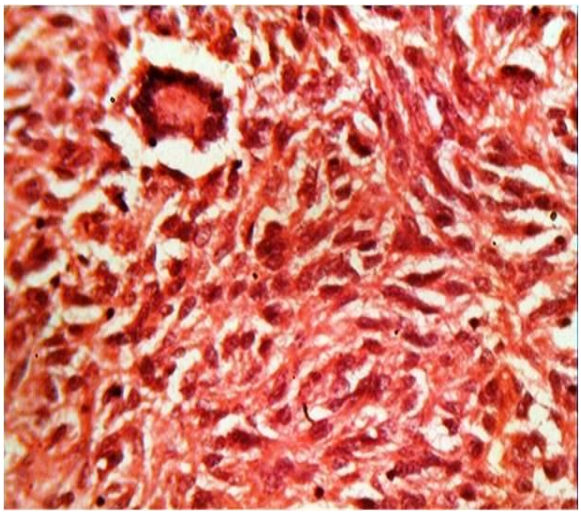

Figure 5: Photomicrograph of occasional tumor giant cells with short, fusiform tumor cells having moderately pleomorphic vesicular nuclei and pale eosinophilic cytoplasm (H & E, x400)

Light Microscopy:

Multiple sections studied showed wall of the colon. A tumor was seen arising from the muscularis propria and infiltrating upto the serosa (figure 3). At places, tumor was invading the mucosa also. The tumor was composed of plump spindle cells arranged in short fascicles, whorls, sheets and nests (figure 4). The tumor cells were short, fusiform having moderately

pleomorphic vesicular nuclei with occasional prominent nucleoli with pale eosinophilic cytoplasm. Occasional tumor giant cells were seen (figure 5). In areas tumor showed epithelioid pattern. Focal areas of hemorrhage and necrosis were noted. Tumor showed moderate (6-8/10hpf) mitotic figures. Serosa showed inflammatory exudates.

Immunohistochemistry (IHC) showed strong and diffuse positivity for C-Kit (CD-117) and focal positivity for smooth muscle actin (SMA).

Final histopathological diagnosis given was malignant gastrointestinal stromal tumor- Grade III with perforation of sigmoid colon with peritonitis.

DISCUSSION

Gastrointestinal Stromal Tumors (GISTs) have only recently become recognized as a distinct pathological entity. The term was first used in 1983 by Mazur and Clark to encompass gastrointestinal and mesenchymal non epithelial neoplasms that lacked the immunohistochemical (IHC) features of schwann cells and did not have the ultrastructural characteristics of smooth muscle cells. [7] The vast majority of GISTs are positive for CD 117 and vimentin in IHC staining. [8] They are different from GI smooth muscle tumors (leiomyoma/leiomyosarcoma) and neural tumors (schwannoma). Tumor was previously described as gastrointestinal autonomic nerve tumor (GANT) is now regarded as variants of GIST. [8, 9]

At present, GISTs may be defined as morphologically spindle cell, epithelioid or occasionally pleomorphic mesenchymal tumors originated from the intestinal cells of Cajal / related stem cells that usually express C- Kit (CD – 177) protein in 95 % of the cases regardless of the site of origin, histological appearance and biological behavior. [10]

GIST usually affects middle aged and older patients with median age of 50-60 years. [2] About 60- 70 % of GISTS occur in the stomach, 20 -30 % in the small intestine and 5 % or less in the colon. The clinical presentation depends on the size and site of the tumor. Small, less than 2 cm GISTs are asymptomatic and incidentally detected at laparotomy or in other related conditions for endoscopy. [2]

Larger tumors present with vague abdominal discomfort, chronic GI bleeding, intestinal obstruction or altered bowel habits. [2] Very large GISTs presenting as externally palpable intra abdominal masses are likely to be malignant. [2] However colonic GIST presented with perforation and peritonitis is extremely rare phenomenon. Standard treatment of GIST is surgical resection. Imatinib mesylate is the standard regimen if the tumor is metastatic or unresectable. [2, 8]

After extensive review, only one case report of spontaneous rupture of GIST in colon was seen in the literature. [4]

In present case, the 70 year old male presented to surgical OPD with chief complaint of pain in the abdomen. On the basis of clinical and radiological examination, emergency laprotomy was carried out and revealed perforation of sigmoid colon with tumor mass protruded through the defect. Histopathological and IHC examination confirms malignant GIST.

GISTs are usually unencapsulated but well circumscribed masses. The cut surface has a whorled fibroid like / softer more fleshy appearance. Large lesions show cystic degeneration or central necrosis. Ulceration of the overlying mucosa is common. The predominant histological pattern seen in 70-80 % of GISTs is of a spindle cell tumor with a fascicular or storiform growth pattern. [10, 11] About 20-30

% of tumors are predominantly composed of large round or polygonal epithelioid cells with abundant eosinophilic to clear cytoplasm. [10, 11] Mixed spindle and epithelioid tumors are common.

In our case, perforated mass in the wall of colon with grey white, fleshy appearance on cut section with predominantly spindle cell tumor histology was noted. IHC showed strong and diffuse positivity for C-Kit (CD-117) & focal positivity for smooth muscle actin.

In our case, size of the tumor was more than 5 cm, high cellularity, with presence of necrosis, C-Kit positivity and mitosis more than 6 /hpf suggest malignant nature of GIST and graded as Grade III.

Differential diagnosis:

GIST has to be distinguished from true smooth muscle tumors and schwannomas of the GIT. The other differentials include intra abdominal fibromatosis, inflammatory fibroid polyp, paragangliomas and metastatic malignant melanoma. Histological examination will easily distinguished GIST from these entities and further supported by IHC.

CONCLUSION

Colonic GISTs are rare clinical entity accounting for only 5% of tumors arising in the digestive tract. GIST should be included in the differential diagnosis of colorectal mass.

Spontaneous rupture is very rare manifestation of a GIST. To the best of our knowledge this is second case report of malignant GIST of sigmoid colon with perforation and peritonitis after Hwangbo Y et al [4] (2011) in the literature.

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