

Brief Communication

A Case of Gastrointestinal stromal tumor (GIST) with review

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A case of GIST

44 years old gentleman presented to surgery department - vague abdominal pain of uncertain duration with abdominal mild distention & discomfort at Tawau general Hospital in Sabah state of Malaysia. During routine physical examination ill-defined mass was palpable in the left upper hypochondriac region for which Barium enema X Ray was requested to rule out splenic flexure colonic mass but it revealed no abnormality. Subsequently Ba-meal follow through was advised to rule out intestinal obstruction and the study showed narrowing of terminal ileum long segment with proximal dilatation of small gut, no other abnormality could be demonstrated. Ultrasonography also showed ill-defined mass in central abdominal cavity. As the patient condition was not improving CT abdomen was also done. CT scan showed ill defined large solid cystic mass mainly occupying the central abdomen merged with the mesentery & gut which was reported as retroperitoneal mesenteric sarcoma. Then exploratory laparotomy was done which demonstrated the irregular large mass lesion mainly involving the small gut (ileal part), mesentery & part of transverse colon. Surgeons attempted to be resect the mass but complete resection was not possible due to diffuse involvement of the tumor. The specimen was sent for histopathology which came out as gastrointestinal autonomic nerve cell sarcoma or gastro intestinal stromal tumor (GIST).

One year later repeat CT abdomen showed residual tumor. Two months later as the condition of patient was deteriorating rapidly repeat CT scan of abdomen was done again. It showed recurrence of huge mass lesion spreading from upper abdominal region to pelvic cavity. Unfortunately patient died a week later.

Discussion:

Gastrointestinal stromal tumors (GISTs) are a subset of GI mesenchymal tumors of varying differentiation¹. Previously, these tumors were classified as GI leiomyomas, leiomyosarcomas, leiomyoblastomas, or schwannomas depending on their histologic findings and apparent origin in the muscularis propria layer of the intestinal wall. GISTs account for approximately 80% of GI mesenchymal tumors.

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Grossly, GISTs are well-demarcated spherical masses that appear to arise from the muscularis propria layer of the GI wall. Intramural in origin, they often project exophytically and/or intraluminally, and they may have overlying mucosal ulceration. Larger GISTs nearly always outgrow their vascular supply, leading to extensive areas of necrosis and hemorrhage. The diameter of GISTs, as a whole, can range from a few millimeters to more than 30 cm. Cytologically, GISTs can be classified into spindle cell GISTs and epithelioid GISTs. Independent of location, most GISTs express the CD34 antigen (70-78%) and the CD117 (72-94%) antigen. The CD34 protein is a hematopoietic progenitor cell antigen that occurs in a variety of mesenchymal tumors. CD117 also is known as the c-kit protein; it is a membrane receptor with a tyrosine kinase component. Mutations in the CD117 gene have been linked to malignant behavior in GISTs^{2,3}. GISTs are rare tumors, constituting less than 3% of all GI malignant neoplasm⁴. On presentation, 41-47% of malignant GISTs are metastatic. GISTs have a unimodal peak incidence in persons aged 40-70 years, but they have a broad distribution. About 50-70% of GISTs occur in the stomach, 33%, in the small bowel, 5-15%, in the procto colon, and only 1-5%, in the esophagus. GISTs are multicentric in fewer than 5% of cases. About 10-30% of GISTs have malignant behavior. GISTs rarely spread to regional lymph nodes (<10%). Its malignancy is manifested by local invasion; distant metastases most commonly involve the liver (50-65%) and peritoneum (21-43%). Only 10% of metastatic lesions occur in the lungs or bones⁵. These tumors have a wide clinical spectrum at presentation. They are often asymptomatic or incidentally detected. If symptomatic, GISTs usually cause symptoms as a result of their size or tendency to ulcerate and bleed. The most common presenting signs and symptoms include abdominal pain, GI bleeding, and a palpable mass. Although these tumors rarely cause obstruction, they can become perforated in as many as 20% of cases.

Radiologic Overview of the Diagnosis⁶

Ultrasonography

On sonograms, larger GISTs appear as complex masses with cystic and solid components, which are consistent with their tendency to necrose. Endoscopic ultrasonography can be valuable in the evaluation of GISTs. The tumors appear as hypoechoic masses that are contiguous with the fourth hypoechoic layer of the GI wall, which corresponds to the muscularis propria.

Computed Tomography

Smaller GISTs appear as smooth, sharply defined intramural masses with homogenous attenuation. Contrast enhancement may be rim like or uniform. Occasionally, dense focal

calcifications are present. Larger GISTs with necrosis appear as heterogeneous masses with enhancing borders of variable thickness and irregular central areas of fluid, air, or oral contrast attenuation that reflect necrosis. Overlying mucosal ulcerations and extension into nearby structures may be present. CT is also sensitive for the detection of metastatic liver, peritoneal, lung, and bone lesions. Liver lesions can be hyper vascular or appear as cystic multi locular lesions with fluid-fluid levels.

MR Imaging

GISTs appear as sharply delineated, heterogeneous masses with cystic and necrotic areas. The masses tend to be isointense relative to skeletal muscle on T1-weighted images and hyper intense on T2-weighted images. Signal intensity voids are present if gas is present within areas of necrotic tumor.

Management and Treatment ⁷

Previously, the only proven treatment was surgical resection. Radiation therapy and chemotherapy have been ineffective. The drug Imatinib mesylate, sold under the trade name Gleevec in the United States, is currently being studied in clinical trials at several institutions. Imatinib mesylate is an inhibitor of tyrosine kinases (KIT receptor and the PDGRF receptors). The drug has proven effective in controlling the disease in early- and late-stage disease. Post treatment, liver lesions become more well defined and cyst like.

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