

ROLE OF MDCT IN GASTROINTESTINAL STROMAL TUMORS WITH PATHOLOGICAL CORRELATION

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ABSTRACT

Gastrointestinal Stromal Tumors (GISTs) are mesenchymal tumors that arise from precursors of the connective tissue cells of the gastrointestinal tract. The aim of this study was to review the role of MDCT in detection and characterization of GISTs with pathological correlation. **MATERIALS AND METHOD:** 22 patients suspected of having GIST were examined using 64 slice MDCT over a period of 2 years. Two radiologists reviewed the CT findings by consensus with respect to lesion size, contour, tumor enhancing pattern, mesenteric fat infiltration, ulceration, calcification, lymphadenopathy, local and distant metastasis. **RESULTS:** In this study 12 patients were confirmed on biopsy as having GIST, 11 patients underwent surgery and were started on Glivec (Imatinib), 2 patients had a recurrence and 1 patient was treated by chemo-embolization for hepatic metastasis. **CONCLUSION:** The increasing recognition of GISTs and prolonged survival has made imaging increasingly important not only for diagnosis but also for monitoring the effects of treatment and detecting tumor progression. CT is the imaging modality of choice for these purposes.

Keywords: GIST, MDCT.

Introduction

Gastrointestinal stromal tumors are rare but, nevertheless the most common mesenchymal tumors of the gastrointestinal tract. GISTs occur most commonly in the stomach and the small bowel. CT is the imaging modality of choice for diagnosing and staging of GISTs and for monitoring the disease during and after treatment. The purpose of our study was to describe the CT findings of GIST and to correlate them with pathological diagnosis.

Materials and Methods

This is a retrospective study conducted at Advanced Radiology Clinic, Karachi. The study was carried out from January 2006 to October 2008. A total of 22 patients were included in this study in which 19 were

male and 03 were female with an age range of 33 to 75 years. Four patients were excluded as they could not be followed up. The patients presented to us with complaints such as abdominal pain, hematemesis, weight loss, vomiting and palpable abdominal mass. Each mass lesion was assessed for the presence of endophytic and exophytic components and areas of low attenuation on CT, the pattern of enhancement on contrast administration was also assessed. The adjacent organs and fat planes were assessed for evidence of invasion.

Image Acquisition

CT was performed on a 64-MDCT scanner (Aquilion 64, Toshiba Medical System Corporation) at our centre. Patients were asked to fast for 6 hours before scanning. Oral and intravenous contrast material was administered and dual phase scanning was performed of the abdomen and pelvis.

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Results

Out of 22 patients, 12 diagnosed as GIST on MDCT were also confirmed on biopsy Table 1. One patient was confirmed as Carcinoid and one of them had an inconclusive result. Five of them also showed associated lymph node enlargement, three of which were confirmed as Lymphoma on biopsy. Eight patients with GIST involved the stomach, one had a mass in the mesentery, two had a growth in the duodenum and one had a mass involving the rectum. All of them

showed a large heterogeneously enhancing well defined soft tissue mass with areas of necrosis in them. There was associated mass effect over the adjacent structures like bowel loops, ureters and vessels with no evidence of their invasion. Eleven of the patients with GIST underwent surgery and were put on Glivec (Imatinib), two patients had a recurrence and one of them had metastatic deposits in the iliac fossa postoperatively. One was treated with TACE for hepatic metastases with inhomogenous uptake of the lipiodol by the mass in the follow up studies.

TABLE 1: CLINICAL AND IMAGING FINDINGS IN PATIENTS WITH BIOPSY PROVEN GIST

Age (yr)	Sex	Presenting symptoms	Site of Tumor	MIDCT Appearance at presentation	MDCT Appearance at Follow-up
33	M	Abdominal pain, weight loss	Duodenum	Mass arising from second part of duodenum; hepatic metastasis	Postop case on Glivec; no recurrence, no change in size of hepatic lesion.
45	M	Indigestion, abdominal pain	Stomach	Large gastric mass at lesser curve; areas of necrosis.	Postoperative GIST on Glivec; no recurrence or metastasis.
45	M	Vague abdominal pain, weight loss	Stomach	Large gastric mass with exophytic component involving body and tail of pancreas; no hepatic or nodal metastasis	Postop case on Glivec; residual mass in tail of pancreas; no recurrence
55	M	Abdominal pain, weight loss	Duodenum	Large duodenal mass; exophytic pancreatic component; no metastasis	Postop on Glivec; No recurrence; no metastasis.
47	M	Weight loss, Constipation, melaena	Rectum	Large rectosigmoid mass with endo- and exophytic components and extending into the retroperitoneum, hepatic metastasis	GIST with surgery of the large rectosigmoid mass and chemo-embolization of hepatic lesions; patchy uptake of lipiodol; no significant change in size of the lesions.
47	M	Abdominal pain, weight loss	Stomach	Gastric fundal mass with large pancreatic component with areas of necrosis; no metastasis	Postop GIST following Whipple's; metastatic deposits in mesentery anterior to IVC.
48	M	Weight loss, melena	Mesentery	Multiple mesenteric masses; areas of necrosis; hepatic metastases	Postop on Glivec; mesenteric recurrence with nodal and hepatic metastases.
52	M	Indigestion, weight loss	Stomach	Large gastric fundal mass; areas of necrosis;	Postop on Glivec; no recurrence; no metastases
56	M	Abdominal pain, weight loss, vomiting	Stomach	Large polypoid gastric mass involving body; presence of air lucencies in it	Biopsy proven GIST; no recurrence; no metastasis
60	M	Abdominal pain, vomiting	Stomach	Large gastric fundal mass; areas of necrosis; no metastasis	Postop on Glivec; no recurrence; no metastasis
70	F	Bloatness, vomiting, weight loss	Stomach	Large gastric mass with exophytic pancreatic component; no vessel involvement	Postop GIST on Glivec; residual mass in tail of pancreas; no recurrence; no metastasis
75	M	Weight loss, abdominal pain	Stomach	Large gastric fundal mass; no metastasis	Postop on Glivec; recurrent mass in stomach; metastatic deposits in both iliac fossae

Note: GIST - gastrointestinal stromal tumor.

Images



Figure 1: 56 year old male with gastric gastrointestinal stromal tumor. Axial and coronal CT scans showing a polypoid gastric mass involving body with presence of air lucencies in it.

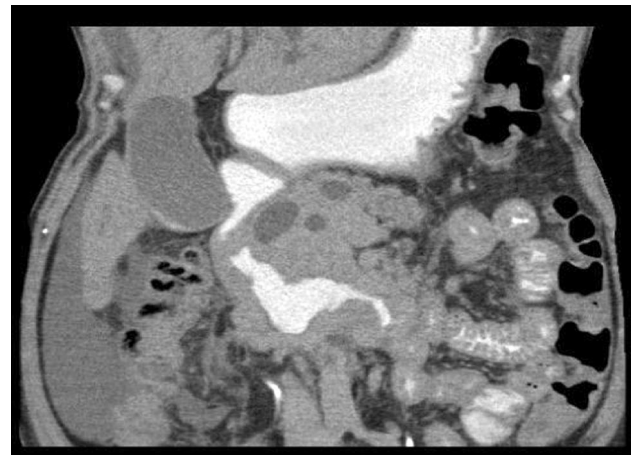
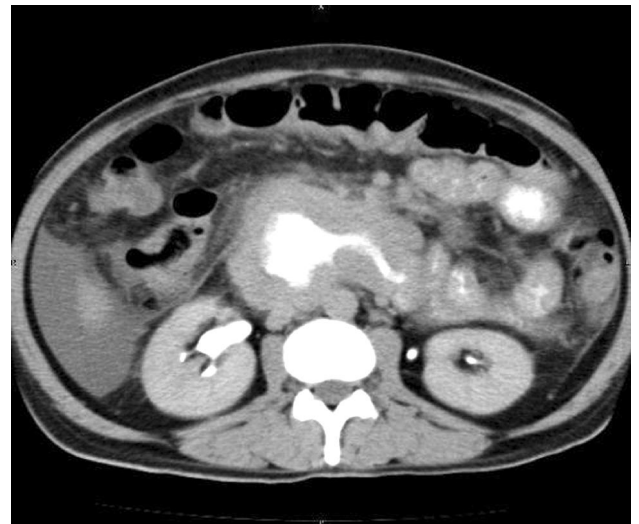


Figure 2: 55 year old male with duodenal gastrointestinal stromal tumor. Axial and coronal CT scans showing a large soft tissue mass involving 2nd and 3rd parts of duodenum causing its narrowing and having an exophytic pancreatic component with dilatation of common bile duct and main pancreatic duct. Ascites is also seen.

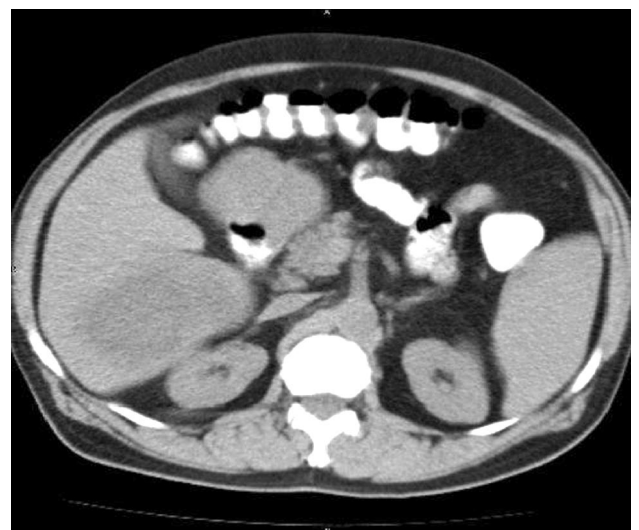


Figure 3: Axial CT image of a 33 year old male with duodenal GIST involving 2nd part of duodenum with metastatic hypodense lesion in the right lobe of liver.



Figure 4: Contrast-enhanced coronal image of a biopsy proven 48 year old male with GIST showing multiple inhomogeneously enhancing mesenteric masses of varying sizes.



Figure 5: 47 year old male with gastrointestinal stromal tumor on Glivec. Axial contrast-enhanced CT scans of mid abdomen showing an inhomogeneously enhancing soft tissue mesenteric mass with considerable reduction in its size after taking Glivec for 5 months

Discussion

Gastrointestinal stromal tumors (GIST) are mesenchymal tumors which typically arise in association with the muscularis propria of gastrointestinal (GI) tract wall.¹ They are found in the stomach (60%), small bowel (30%), the colon and rectum (5%), esophagus (<5%) and mesentery. Most GISTs occur typically between the ages of 50-60.² Patients may present with gastrointestinal bleeding, anemia, abdominal pain, dyspepsia or a palpable abdominal mass.³ In a study by Nishida et al, of 271 patients with stromal tumors 2/3 had symptoms that correlated with tumors size. Tumors larger than 3 cm were more likely to demonstrate necrosis than tumors less than 3 cm.⁴ Histopathologically most spindle cell tumors in the gastrointestinal tract are GIST. GISTs are composed of spindle (70%) or epithelioid (30%) cells.⁵ These tumors are both phenotypically and genotypically different from true leiomyomas and usually express CD34, a hematopoietic progenitor cell antigen. CD34, however, is also present in a wide variety of fibroblastic and endothelial cell tumors⁶ GISTs differ clinically and pathogenetically from true leiomyosarcomas (very rare in the GI tract) and leiomyomas. The latter occur in the GI tract, predominantly in the esophagus (intramural tumors) and the colon and rectum (muscularis mucosae tumors).⁷

The vast majority of GISTs express a mutant form of c-kit (CD117).⁸⁻¹⁰ The clinicopathological features of GISTs with or without c-kit mutations are markedly different. Therefore, GISTs may be divided into four major categories based on histochemical and genetic data: myogenic tumors; neurogenic tumors; GISTs with c-kit mutation; and GISTs without c-kit mutation.¹¹ The symptoms and signs of GISTs are not disease-specific, so that about 50% of them already have metastases at the time of diagnosis, usually to the liver or the peritoneum.¹² There are various factors that have been correlated with aggressive behavior such as large tumor size, presence of metastasis at surgery, high mitotic counts, high proliferation indices and tumor necrosis. The liver is the most common site of metastases followed by peritoneum. Ascites is very rarely seen. Malignant stromal tumors can invade adjacent organs and can metastasize hematogenously,

usually to the lung or liver. Metastatic lesions may also appear low in attenuation due to necrosis¹³ They rarely obstruct viscera, despite their large size and propensity to metastasize to the liver and peritoneum.¹⁴ CT is considered to be the imaging modality of choice for the detection, staging, surgical planning and follow-up of patients with GIST. New MDCT with the ability to obtain thin collimation and high resolution allows for detection of tumors measuring 1.0cm and above. As all tumors 2 cm or greater in size are detected with MDCT, these findings suggest that MDCT can be useful in screening symptomatic patients suspected of having GISTs.⁴ In general, contrast enhanced CT is as reliable as FDG PET in the evaluation of treatment responses. FDG PET is indicated whenever CT findings are inconsistent or inconclusive.

On contrast enhanced CT, localized primary GISTs are typically exophytic, large, hypervascular masses. Small tumors tend to appear homogeneous. The larger tumors (>6cm) frequently show central areas of necrosis or hemorrhage. Central gas and mural calcification are uncommon findings. The majority show peripheral enhancement. They may appear as intramural masses or intraluminal polyps, and may show extension into adjacent mesentery. Encasement of adjacent small bowel, colon, and bladder can be seen. Lymphoma may share many features of small intestinal GISTs, but is notable for lymphadenopathy.¹⁵

Prediction of clinical behavior in this group of tumors is notoriously difficult, and the same criteria for malignancy do not necessarily apply to stromal tumors from different sites within the gastrointestinal tract.¹⁶ Goldblum and Brainard et al concluded in a study that based on light microscopic features alone, benign and malignant duodenal, jejunal and ileal stromal tumors can be separated from each other.¹⁷⁻¹⁸ In the largest pathologic series of anorectal gastrointestinal stromal tumors with long-term follow up, Miettinen et al. established that 70% of rectal gastrointestinal stromal tumors less than or equal to 2 cm in maximal dimension and having no more than five mitoses per 50 high-power fields showed malignant behavior and were associated with significant tumor-related mortality rates.¹⁹⁻²⁰

There is no general agreement on the prognosis of GISTs. Surgical resection is the conventional therapy for GISTs. However, overall prognosis of patients with

GISTs treated with surgery alone is discouraging. Rossi et al report that 10 out of 18 patients in their series developed recurrent disease following radical surgery.²¹ Even in patients who undergo surgery to treat relapsed disease, median survival drops to 15 months, compared to 50 months in primary cases. Most gastrointestinal stromal tumors (GISTs) have activating mutations in the KIT receptor tyrosine kinase, and most patients with GISTs respond well to Gleevec, which inhibits KIT kinase activity.¹⁵ Gleevec has become the standard chemotherapy for metastatic or unresectable tumors.²² When the tumors respond to treatment, the changes in tumor size may initially vary; however, GISTs typically become homogenous and hypoattenuating, with disappearance of enhancing tumor nodules and tumor vessels in the early post-treatment period?. Development of a nodule within treated tumor indicates recurrence regardless of changes in tumor size.¹⁵

Hepatic metastases from gastrointestinal stromal tumors that respond to treatment with STI-571 can appear as near-cystic components with well-defined borders on contrast-enhanced CT. These metastases resemble simple cysts, but density measurements may differentiate them from one another.²³

Conclusions

Gastrointestinal stromal tumors (GIST) are mesenchymal tumors that typically arise in association with the muscularis propria of gastrointestinal (GI) tract wall. CT can be a useful modality for the diagnosis, staging and follow-up of patients with GIST. It is important for the radiologist to be familiar with proper CT technique for imaging these patients as well as the variety of CT appearance of the primary tumors and metastases.

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