

**Case Report****The gist of GISTS – A radiological case report****Dr. Sumit Sharma, Dr. S. Shrinivasan, Dr. R. Chidambaram**Department of Radiodiagnosis, Sri Lakshmi Narayana Institute of Medical Sciences, (Affiliated to Bharath University)  
Osudu, Puducherry, India**\*Corresponding author**

Dr. S. Shrinivasan

Email: [drshrinivas@gmail.com](mailto:drshrinivas@gmail.com)

**Abstract:** Gastrointestinal stromal tumours (GISTs) constitute a group of smooth muscle mesenchymal gastrointestinal tract tumours. These are found to originate from the interstitial cells of Cajal 2-3, with 95% staining positive for CD117 (c-KIT) and 70% for CD34. The former is highly responsive to the chemotherapeutic agent ST-571 (Imatinib). The lesion morphology varies according to location but usually they appear as rounded irregular soft tissue protrusions, arising from the wall of a hollow viscus (mostly stomach) projecting into the lumen. We report here a case of a fifty-year-old male with complaints of a vague abdominal pain with nausea and occasional bouts of vomiting and present the imaging findings.

**Keywords:** GISTS, Imatinib, Imaging findings

**INTRODUCTION**

Gastrointestinal stromal tumours (GISTs) are the most common mesenchymal tumours of the gastrointestinal tract accounting for approximately 5% of all sarcomas. GISTs are mostly seen in older patients, rarely seen before the age of 40. GISTs may present with dysphagia, gastrointestinal bleeding, or metastases (mainly in the liver). GISTs arise in the smooth muscle pacemaker interstitial cell of Cajal, or similar cells. They are defined as tumors whose behaviour is determined by mutations in the KIT gene (85%), PDGFRA gene (10%), or BRAF kinase (rare) [1]. They respond remarkably well to a chemotherapeutic agent ST-571 (Imatinib). The definitive diagnosis is made with a biopsy, which can be obtained endoscopically, percutaneously with CT or ultrasound guidance or at the time of surgery. The role of radiologic imaging is to locate the lesion, determine the morphology, evaluate for signs of invasion and detect metastasis. Features of GIST vary depending on tumour size and organ of origin. The diameter can range from a few millimetres to more than 30 cm. Larger tumours usually because symptoms in contrast to those found incidentally which tend to be smaller and have better prognosis [2, 3]. Large tumours tend to exhibit

malignant behaviour but small GISTs may also demonstrate a clinically aggressive behaviour [4]. All GIST tumors are now considered to have malignant potential, and no GIST tumor can be definitively classified as benign [5]. We report here a case of a fifty-year-old male with complaints of a vague abdominal pain with nausea and occasional bouts of vomiting and present the imaging findings.

**CASE REPORT**

A fifty-year-old male with complaints of a vague abdominal pain with nausea and occasional bouts of vomiting on USG was found to have a large well-defined intraluminal hypoechoic mass lesion with irregular margins measuring approximately 13.8 X 11.1 cms arising from the submucosa of the fundus of the stomach. On Doppler, the well-circumscribed mass was found to have a few peripheral vessels. On CT, a large well defined intraluminal heterogeneous mass lesion (measuring 14 X 11 cms) with central areas of low attenuation (suggestive of necrosis) and irregular asymmetrical circumferential wall thickening was seen arising from the submucosa of the fundic region, causing elevation of the left dome of the diaphragm and displacement of the left kidney and spleen peripherally.



**Fig-1:** Ultrasonography images (a) (b) shows an irregular intraluminal and exophytic hypoechoic mass arising from the gastric fundus, measuring approx. 13.8 X 11.1 cms. (c) Doppler image shows few peripheral vessels around the mass



**Fig-2:** Axial CT images (a) (b) (c) shows a large well defined intraluminal and exophytic heterogeneous mass lesion (blue arrow) with central areas of low attenuation (red arrow), irregular circumferential wall thickening (white arrow) arising from the submucosa of the fundic region, causing elevation of the left dome of the diaphragm and displacement of the left kidney and spleen peripherally (Yellow arrow).

**DISCUSSION**

The purpose of radiologic imaging is to locate the lesion, evaluate for signs of invasion and detect metastasis. Large tumors tend to exhibit malignant behavior but small GISTs may also demonstrate clinically aggressive behavior [4]. Plain radiographs are not very helpful in the evaluation of GISTs. On abdominal x-ray, gastric GISTs may appear as a radiopaque mass altering the contour of the gastric air shadow. If cavitations are present, plain radiographs will show collections of gas within the tumor [6].

Barium fluoroscopy and CT are commonly used to evaluate the patient with abdominal complaints. Barium swallows show abnormalities in 80% of GIST cases [4]. However, those GISTs which may be located entirely outside the lumen, will not be differentiated by a barium swallow. Even when the barium swallow is abnormal, a CT/MRI scan must be taken since it is impossible to evaluate abdominal cavities and organs with a barium study alone. In a CT scan, abnormalities

may be seen in 87% of patients and it should be made with both oral and intravenous contrast [4].

Preferred imaging modalities in the evaluation of GISTs are CT, MRI [7] and endoscopic ultrasound. Advantages of CT include its ability to demonstrate evidence of nearby organ invasion, necrosis, ascites, and metastases.

Since GISTs arise from the bowel layer called muscularis propria, small GIST imaging usually suggests a submucosal origin. In barium swallow studies, these GISTs present with smooth borders forming right or obtuse angles with the nearby mucosal wall, as seen with any other intramural mass. The mucosal surface is usually intact with areas of ulceration, which are generally present in half of GISTs.

The tumor may grow exophytically or intraluminally but they most commonly grow exophytically such that the bulk of the tumor protrudes into the abdominal cavity. If the tumor outstrips its

blood supply, it can necrose internally, creating a fluid-filled cavity with hemorrhage and cavitations that can eventually ulcerate and communicate into the lumen of the bowel. In that scenario, barium swallow may show an air /air-fluid level or oral contrast media accumulation within these areas [4, 8]. Mucosal ulcerations may also be present. In contrast enhanced CT images, large GISTs appear as heterogeneous masses due to areas of tumor cells surrounding hemorrhage, necrosis and cysts, which is radiographically seen as a peripheral enhancement pattern with a low attenuation center [3].

In our case, the lesion shows asymmetrical circumferential wall thickening with both exophytic and intraluminal component showing necrotic areas. The lesion displaces left kidney and spleen peripherally. There is no evidence of adjacent nodal and liver metastases. Endoscopically biopsy was taken from the intraluminal component which shows features of GIST. Malignancy is characterized by local invasion and metastases, usually to the liver, omentum and peritoneum. In contrast to gastric adenocarcinoma or gastric lymphoma, malignant lymphadenopathy (enlarged lymph nodes) is uncommon (<10%) and thus imaging usually shows absence of lymph node enlargement [3]. Malignant behavior is less commonly seen in gastric GISTs, with a ratio of behaviorally benign to overtly malignant of 3-5:1 [3]. Definitive diagnosis must be made immunochemically.

Surgical en-bloc resection is the primary mode of therapy for GISTs [9]. The management of malignant GISTs has been revolutionized by the development of Imatinib which is a therapeutic agent that targets a specific abnormal intra-cellular signaling molecule. An effective management of patients with these tumors requires regular imaging assessment for which CT has conventionally been the method of choice [10].

## CONCLUSION

GISTs are incidentally identified on imaging for other indications but mostly present with dysphagia. The vast majority of GISTs are sporadic. However, they occasionally occur in the setting of a recognized syndrome like Carney triad (gastric GIST, pulmonary chondroma and extra-adrenal paraganglioma) and Neurofibromatosis type 1 (NF1). They can be managed well with Imatinib therapy and en-bloc resection. Therefore, it is important to diagnose these tumors at the earliest for a better prognosis and treatment.

## REFERENCES

1. Demetri, G; DeVita, L; Lawrence, TS; Rosenberg, SA, editors "Chapter 87". DeVita, Hellman, and Rosenberg's Cancer: Principles and Practice of Oncology (9th Ed.). 2011.

2. Burkill GJ, Badran M, Al-Muderis O, Meirion Thomas J, Judson IR, Fisher C, Moskovic EC. Malignant gastrointestinal stromal tumor: distribution, imaging features, and pattern of metastatic spread 1. *Radiology*. 2003 Feb; 226(2):527-32.
3. Hersh MR, Choi J, Garrett C, Clark R. Imaging Gastrointestinal Stromal Tumors". *Cancer Control*. 2005; 12(2):111-115.
4. Pithorecky I, Cheney RT, Kraybill WG, Gibbs JF. Gastrointestinal stromal tumors: current diagnosis, biologic behavior, and management. *Annals of Surgical Oncology*. 2000 Oct 1; 7(9):705-12.
5. Raut CP, DeMatteo RP. Evidence-guided surgical management of GIST: beyond a simple case of benign and malignant. *Annals of Surgical Oncology*. 2008 May 1; 15(5):1542-3.
6. Shojaku H, Futatsuya R, Seto H, Tajika S, Matsunou H. Malignant gastrointestinal stromal tumor of the small intestine: radiologic-pathologic correlation. *Radiation medicine*. 1996 Dec; 15(3):189-92.
7. NCCN Clinical Practice Guidelines in Oncology Soft Tissue Sarcomas, version 3.2012. National Comprehensive Cancer Network.
8. Lehnert T. Gastrointestinal sarcoma (GIST): a review of surgical management. In *Annales chirurgiae et gynaecologiae*, 1998; 87(4):297-305.
9. Maki RG. Gastrointestinal stromal tumors (GIST) and their management. *Gastrointestinal cancer research: GCR*. 2007 Jul; 1(4 Suppl 2):S81.
10. King DM. The radiology of gastrointestinal stromal tumours (GIST). *Cancer Imaging*. 2005 Dec 15; 5(1):150-6.