An Epigastric Lump: An Innuendo of a Pedunculated Giant Gastric GIST. An Exotic Presentation.

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Introduction
Gist are apparently a rare group of neoplasms with the incidence ranging from 6.5 to 14.5 / million per year. ¹

Though they are the most common (80%) of the mesenchymal tumor of the alimentary canal ².

It accounts for 1 % of all the gastrointestinal tumours and about 5% of all sarcomas. ³

The incidence of GIST has been historically underestimated prior to the introduction of CD117 staining because of lack of proper diagnostic parameters.

After the advent of CD117 staining (c-kit) for GIST, the incidence of reporting of GIST has increased.

GISTs are mostly found in stomach (55.6-59%) and secondly in small intestine (primarily jejunum/ileum, 25-31.8%) followed by colorectal (3-6.0%), other sites (3-5.5%), and the esophagus (0.7%). ⁴

Only a small number of cases (<1%) have been reported in the appendix.

On rare occasions, GISTs develop outside the gastrointestinal tract, in the mesentery, omentum, or retroperitoneum.

Most patients with GISTs are asymptomatic although patients with advanced disease may present with symptoms of a mass lesion, abdominal pain, or bleeding.

At least 10 to 30% of GISTs are discovered incidentally during laparotomy, endoscopy, or other imaging studies, with 15% to 50% of GISTs presenting with metastatic disease.⁵

Here we present a case report of a Giant gastric GIST presenting as an epigastric lump, which is a rare presentation.

Case Report
A 40 year old male, came to general surgery OPD with complaints of an abdominal lump present in the upper quadrant of the abdomen, gradually increasing in size over the past 5 years. Initially it was roughly 3 x 2 cm in size as quoted by the patient and later it attained its presenting size of roughly 10 x 7 cm. Patient complained of an intermittent dull aching pain, early satiety. Patient had no other presenting complaint.

Patient had no history of weight loss, melena, haematemesis, change in bowel or bladder habits.

On examination, there was presence of a firm mass of about 10 x 7 cm, in epigastric region, extending to left hypochondrial region, the mass moved with respiration, was non tender, non-pulsatile, lower margin of the lump was well defined, the upper margin could not be assessed.

The swelling had side to side mobility.

There were no other scars, sinuses, fistulas that were present.
No supraclavicular swelling, no testicular swelling, no visible peristalsis, hernia sites were normal.

Patient had no significant past history of disease. No similar history of disease in the family.

On blood investigations were within normal limits and viral serology for HIV, HBSAG HCV was negative.

On imaging, Sonography of abdomen revealed a poorly echogenic lesion measuring 93 x 67 mm, predominantly solid in nature seen in epigastric region under the abdominal wall most likely arising from pancreas.

Due to difficulty in assessing the true nature of the lesion with USG, A sonographic guided FNAC was performed which suggested an adenomatous lesion, probably benign in nature.

A Contrast enhanced CT was performed which revealed a smooth margin enhancing attenuating lesion predominantly solid in nature measuring 85x75 mm in size in epigastric region abutting the posterior wall of stomach. Intralesional calcification was also noted.

CECT was not able to point to an exact diagnosis and hence patient was posted for an exploratory laparotomy.

Patient was explored by a midline abdominal incision, intra-operatively the entire abdominal cavity was inspected, a bulge was noticed inferior to the greater curvature of stomach covered by the gastro-colic ligament, gastro colic ligament was divided to assess the lesser sac, upon dividing the ligament a 12 x 10 x 7cm lobulated mass with few areas of necrosis was observed, the mass was adherent to the posterior wall of stomach with a small peduncle.

The mass was mobilised and excised with the peduncle, hemostasis was achieved, the gastrocolic ligament was sutured using 2'0 mersilk. The rest of the abdominal cavity was inspected and there were no signs of metastasis. Liver and spleen were specially unremarkable.

The specimen for sent for histopathological examination. Which revealed a gross specimen which was greyish white lobulated soft to firm tissue piece measuring about 12.5 x 10 x 7.0 cm. Cut surface was greyish white with cystic areas.

Microscopic findings were suggestive of sections showing interlacing fascicles of slender spindle shaped cells having slender nuclei and eosinophilic cytoplasm. Focal areas showed whorling and storiform pattern. No mitosis was seen, hence a finding suggestive of benign spindle cell lesion—most probably benign gastrointestinal stromal tumor was made.

On immunohistochemistry tissue was found to be CD117 positive and CD34 positive and S100 negative.

Based upon histology and IHC a diagnosis of GIST was confirmed that too a benign lesion due to well differentiation and no mitosis observed.

Patient was discharged on 10th post operative day on imitanib mesylate 400mg /day and is under regular follow-up.

Discussion

Gist as discussed earlier are rare tumors, which are often diagnosed incidentally are rare tumors, and gastric GIST though the most common (70%) of all the GIST site. Mostly are asymptomatic and if symptomatic usually present as abdominal pain or haemorrhage. 6

Our case in which GIST presented as an epigastric lump is a rare entity as in concurrence with Sachin palit et al, who in 2011 presented a paper on giant GIST presenting as a palpable abdominal mass an unusual presentation in which the quoted the rarity of such a presentation.

Further in our case it was observed that the GIST was a pedunculated GIST arising from the posterior wall of stomach, completely extra-luminal, which is again a rare incidence.
The incidence of GIST tumors are on the rise because of better diagnostic modalities that are present, since the advent of a specific classification and it is important to identify GIST as these tumors if caught early have a good prognosis due to availability of different treatment modalities.

Since the advent of tyrosine kinase inhibitors in the treatment of GIST tumors and the presence of specific markers like CD 117, CD 34, PDGFRα1, BRAF for diagnosis of GIST have made the prognosis of the disease better due to early identification of the disease.

It also necessary to keep in mind the unusual presentation of gist as an abdominal lump especially a giant abdominal lump as such patients might seem to be incurable but it has been observed that if diagnosed as GIST then these tumors can be downstaged with the use of tyrosine kinase inhibitors and can also be converted from an unresectable tumor to resectable tumor and thus the disease can controlled.

The use of neoadjuvant therapy also decreases the operative risks involved with handling a giant tumor plus improves the patient outcome.

References

3. Hirota , Isozaki K et al , Gain of function mutations of C-Kit in Human gastrointestinal stromal tumors science 1998, 577-80
6. Sachin patil et al – Giant gastrointestinal stromal tumors presenting as a palpable abdominal mass an unusual presentation.

Figure