A rare case of giant Gastrointestinal Stromal Tumour (GIST) of the lesser curvature of the stomach

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Introduction

Gastrointestinal stromal tumours (GIST) are non-epithelial malignant tumours of the gastrointestinal (GI) tract that arise from the interstitial cells of Cajal and make up 1–3% of all GI cancers (1). The stomach is the most frequent site of origin, accounting for 60% of GISTs, followed by the jejunum and ileum (30%), duodenum (4-5%), rectum (4%), colon and appendix (1-2%), oesophagus (<1%), and rarely present as extra-gastrointestinal tumours (1). While the majority of GISTs are diagnosed at a smaller size, giant GISTs, which are more than 20 cm, are rare, being less than 5% of all gastric GISTs (2).

GISTs arise from activating mutations in the KIT or, less commonly, PDGFRA proto-oncogenes, resulting in uncontrolled proliferation of the interstitial cells of Cajal or their precursors. Giant gastric GISTs remain asymptomatic for extended periods owing to their slow growth, presenting only when mass effects become prominent; unlike small GISTs, which often present with gastro-intestinal bleeding and anaemia secondary to mucosal ulceration, giant GISTs typically manifest with abdominal distention, discomfort, a palpable mass, or weight loss (2). Diagnosis relies primarily on contrast-enhanced computed tomography, with histopathological examination and immunohistochemistry (positivity for CD117 and/ or DOG1) required for confirmation. Prognosis depends on tumour size, mitotic index and anatomical location, which together determine the risk of disease progression.

The AFIP/ Lasota-Miettinen classification is a risk-stratification system that categorises GISTs into very low, low, intermediate, and high-risk groups for disease progression based on tumour size, mitotic count and primary site, and is used to guide prognosis and the need for adjuvant therapy (1).

We report a case of giant (22 cm) gastric epithelioid GIST arising from the lesser curvature, successfully managed with surgical resection.

Case presentation

A 47-year-old previously healthy female presented with progressive abdominal distention, upper abdominal pain and discomfort, and early satiety for six months. She had lost 5 kg of weight during this period. She denied vomiting, melaena, or haematemesis. Physical examination revealed a firm, smooth-surfaced, non-tender mass occupying the epigastrium and left upper quadrant. There was no hepatomegaly, free fluid or lymphadenopathy.

Both haematological and serological investigations were within normal limits. Initial ultrasonography revealed a heterogeneous mass with internal vascularity occupying the whole upper and mid abdomen. The contrast-enhanced computed tomography (CECT) showed a 21 cm × 18 cm × 14 cm sized heterogeneous lesion with well-defined margins extending from the upper border of L1 to the lower border of L5, suggestive of a GIST arising from the lesser curvature of the stomach (Figure 1). There was no local infiltration or para-aortic or pelvic lymphadenopathy. Upper gastrointestinal

endoscopy revealed an extrinsic compression of the gastric lumen by a smooth, bulging submucosal lesion with intact mucosa (Figure 2) .

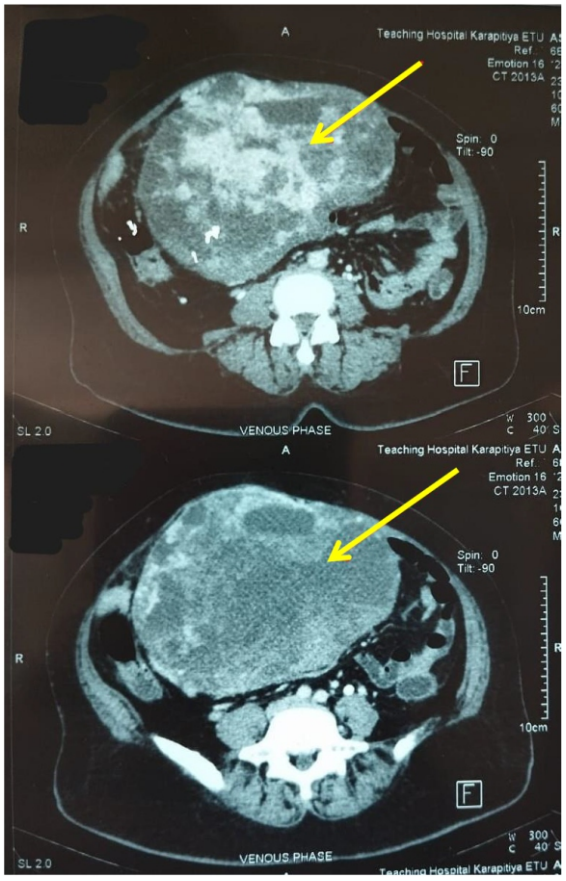

Figure 1: CECT image showing large heterogenous lesion with well-defined margins (yellow arrow)

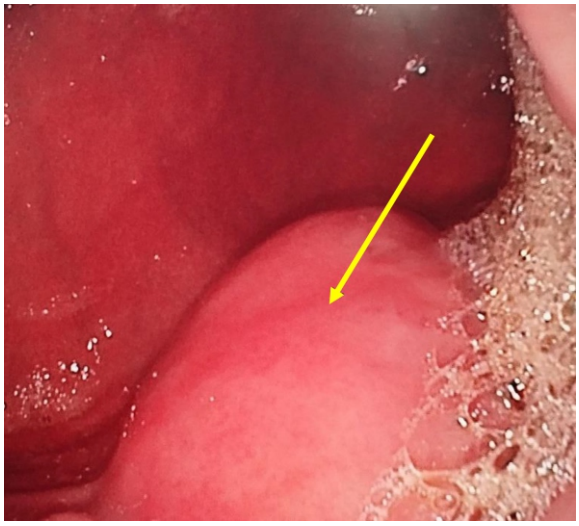

Figure 2: Endoscopic view of the GIST (yellow arrow) from inside of stomach

Due to the large size of the tumour, open surgery with a midline incisional access was performed. Upon opening the abdominal cavity, a large lobulated mass arising from the lesser curvature of the stomach, adherent to the transverse mesocolon and gastroepiploic vessels, was observed (Figure 3 & 4).

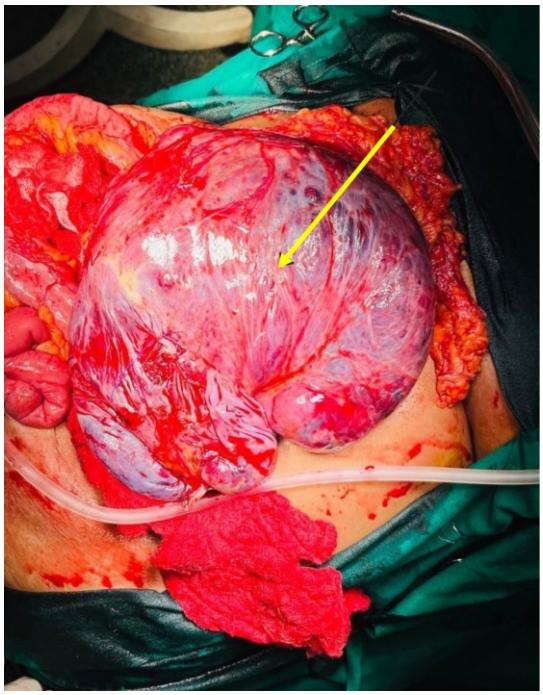

Figure 3: Intra-operative view of large lobulated tumour (yellow arrow) arising from the lesser curvature of the stomach

A wedge resection of the stomach was performed, incorporating the tumour. The adherent portions of the mesocolon were carefully dissected and separated, preventing tumour capsule rupture. The gastric defect was closed in two layers (Figure 5). Her postoperative period was uneventful and was discharged on post - operative day five.

Macroscopic examination showed a tumour measured as 22 cm × 18 cm × 16 cm with no capsular disruption. The cut surface showed a heterogeneous texture with solid and cystic components.

Microscopy revealed an epithelioid GIST of the stomach with high-risk (86%) of progressive disease according to AFIP/Lasota-Miettinen classification (Figure 6). Immunohistochemistry analysis showed diffusely positivity for CD117 and DOG1 and negativity for SMA and CD34.

The patient was referred to medical oncology to start on adjuvant therapy, and a follow-up was arranged.

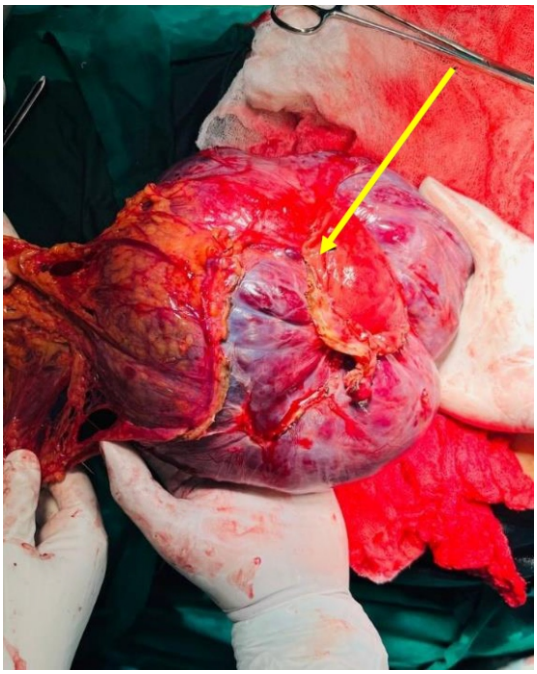

Figure 4: Intraoperative photograph of the large, lobulated gastric GIST held exteriorized during resection (yellow arrow)

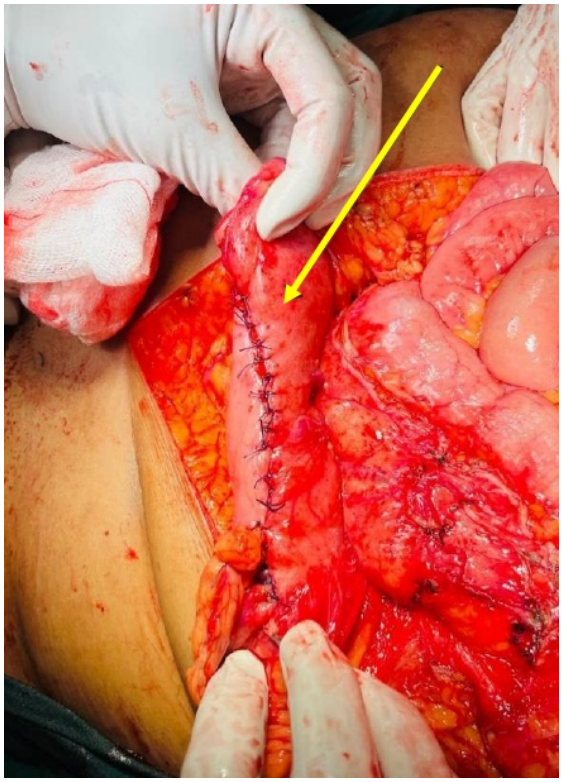

Figure 5: After wedge resection of the stomach with tumour, the gastric defect was closed in two layers (yellow arrow)

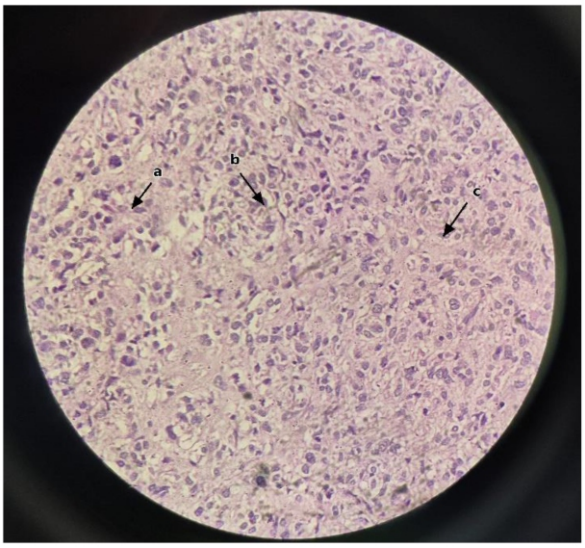

Figure 6: High power microscopy (H&E) image showing scanty stroma (a) with enlarged hyperchromatic nuclei (b) and sheets of epithelioid neoplastic cells (c)

Discussion

Most giant gastric GISTs (60-70%) arise from the fundus or greater curvature, making lesser curvature tumours relatively uncommon (3). CECT is the first choice of imaging modality to assess the primary lesion and look for metastasis. A preoperative biopsy only necessary if there is a suspicion of GIST or surgical resection is impossible (4). Due to the suboptimal accuracy of conventional endoscopic forceps biopsy and the risk of peritoneal seeding and tumour rupture in percutaneous biopsy, endoscopic ultrasound-guided fine needle aspiration (EUS-FNA) is considered a safe and accurate method of biopsy (5).

Complete surgical resection with R0 (micros--copically margin-negative) dissection with a 1 cm tumour margin without rupture is recommended for localised GIST. Endoscopic resection with complete excision without rupture is an alternative for the GISTs <2 cm in size with pre-operatively confirmed GIST (4). Adjuvant therapy with Imatinib 400 mg daily is recommended for patients at high-risk and patients who had tumour rupture. Genotyping should be done prior to treating metastatic patients. First-line treatment is with imatinib. Sunitinib, and regorafenib can be

used in patients with disease progression (4). Debulking surgery with R0 or R1 (microscopic residual disease) surgery can be decided for selected patients following initial imatinib therapy (4).

This case highlights that GIST should remain a key differential diagnosis in patients presenting with a large abdominal mass, even in the absence of classical gastrointestinal bleeding. Although the lesser curvature is an uncommon site for giant gastric GISTs, complete surgical resection with an intact tumour capsule remains feasible and should be the primary goal, as capsule rupture worsens prognosis by increasing the risk of peritoneal dissemination. A multidisciplinary approach involving careful preoperative imaging, meticulous surgical technique, and risk-stratified adjuvant therapy is essential to optimise outcomes in these rare but manageable tumours.

Conclusions

Giant gastric GISTs are a rare clinical entity that often present late due to slow, expansive growth. A precise diagnosis relies on imaging and histopathology (including immunohistochemistry). Complete surgical resection without rupture remains the key to curative management, followed by adjuvant imatinib for high-risk patients.

Informed written consent has been obtained from the patient to publish this case report with photographs.

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