CASE REPORT

A patient with a 9.6-kg gastric gastrointestinal stromal tumour managed at a tertiary referral hospital in Ndola, Zambia

Seke M.E. Kazuma, Bright Chirengendure, Joseph Musowoya, Royd Ngoma, Boniface Kaela, Simunyama Luyando, Kamui Mundia, Elijah Mwale, Matilda Kapompo, Martha Chooka, Rodgers Ngo'mbe

Department of Surgery, Ndola Teaching Hospital, Ndola, Zambia

Correspondence: Dr Seke Manase Ephraim Kazuma (sekekazuma@gmail.com)

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Abstract

Gastrointestinal stromal tumours (GISTs) constitute 1% to 3% of gastrointestinal tract tumours and 5% to 6% of sarcomas, representing the most prevalent subset of mesenchymal tumours within and beyond the gastrointestinal system. GISTs have an annual global incidence of 10 to 20 million cases. The carcinogenesis of GISTs originates from mutations in the *KIT* (also known as *CD117*) and PDGFA (platelet-derived growth factor A) genes within the interstitial cells of Cajal, resulting in the activation of tyrosine kinase receptors. These tumours are typically asymptomatic or incidentally identified during surgery or imaging studies. Diagnosis is confirmed through immunohistochemical staining with a positive *KIT* (*CD117*) marker. For resectable GISTs and those with limited metastases amenable to resection (liver or peritoneal), achieving surgical resection with clear margins is imperative. This report discusses the management of a patient at Ndola Teaching Hospital with a preoperative diagnosis of an ovarian tumour, which intraoperatively was identified as a gastric tumour.

Keywords: gastrointestinal stromal tumours, gastric GISTs, sarcomas, mesenchymal tumours, gastrectomy, tyrosine kinase inhibitors, imatinib, Zambia

Introduction

Gastrointestinal stromal tumours (GISTs) account for 1% to 3% of tumours of the gastrointestinal tract and 5% to 6% of sarcomas; they are the most common subset of mesenchymal tumours within and outside the gastrointestinal tract.[1],[2] GISTs have an annual incidence of 10 to 20 cases per million per year worldwide.[3] These tumours typically originate in the submucosa of the gastrointestinal tract but can also develop extraviscerally from the mesentery and greater omentum.[4] The carcinogenesis of GISTs occurs in the interstitial cells of Cajal, in the myenteric plexus of the gastrointestinal tract, due to mutations in the *KIT* (also known as *CD117*) and *PDGFA* genes, leading to the activation of tyrosine kinase receptors.[5]

GISTs are predominantly asymptomatic and are often incidentally diagnosed during surgery or based on imaging.[6] They can arise anywhere within the gastrointestinal tract: 70% are in the stomach, 25% are in the small intestine, 5% to 10% are in the colon and rectum, 5% are in the oesophagus, and 10% are extravisceral.[7] GISTs can manifest at any age, with the median age at diagnosis being 60 years.[8]

Ninety-five per cent of GISTs are positive for CD117, and the diagnosis is confirmed based on positive immunohistochemical staining for KIT (CD117).[9] Surgical resection with clear margins is essential for resectable GISTs and for GISTs with resectable oligometastasis (liver or peritoneal metastasis).[10]

We report the management of a patient who presented to Ndola Teaching Hospital in Ndola City, Zambia, a resource-limited setting, with an abdominal mass subsequently identified as a gastric GIST on histopathologic examination.

Case presentation

A 53-year-old woman was referred to the gynaecological department at our facility with a 4-month history of a slow-growing abdominal mass associated with abdominal fullness, poor appetite, and weight loss. She denied any changes in bowel or urinary habits and had no symptoms of recurrent hot flashes, night sweats, or jaundice. Her systemic review was unremarkable. Her past medical history included chronic anaemia treated with haematinics; however, she had no comorbidities, such as diabetes mellitus, hypertension,

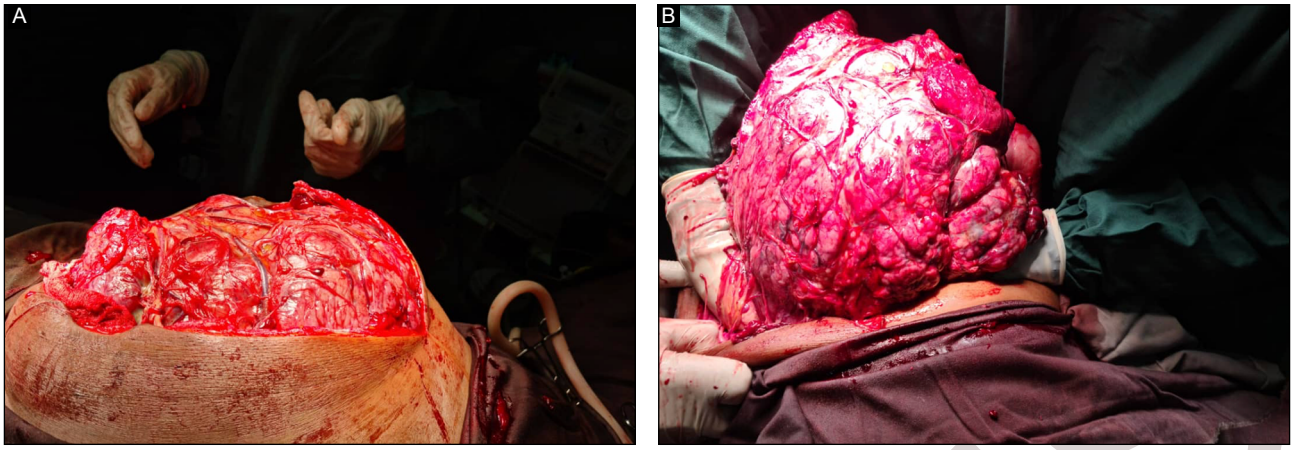

Figure 1. (A) Abdominal tumour found by gynaecologist at laparotomy; (B) tumour partially dissected—noted to be covered in omentum and originating from the distal greater curvature of the stomach

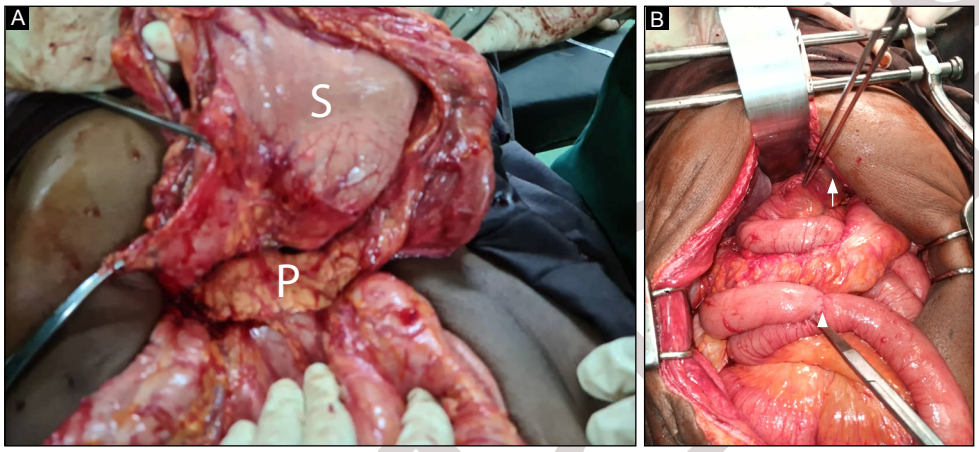

Figure 2. Intraoperative photographs: (A) after tumour excision and total omentectomy, the distal stomach, S, before distal gastrectomy (pancreas, P, indicated for orientation); (B) roux-en-Y end-to-side reconstruction to establish gastrointestinal continuity (arrow: end-to-side distal gastrectomy and proximal jejunum; arrowhead: end-to-side proximal jejunum and distal jejunum)

or HIV, and she had no prior hospital admissions. She was receiving support with haematinics from the referring hospital. The patient was 3 years postmenopausal and had not undergone any cervical cancer screening. She denied any history of smoking or alcohol consumption.

On examination at presentation, she was in good general condition, with no pallor, jaundice, or cyanosis. Her abdomen was globally distended, with a palpable mass extending across all 4 quadrants, measuring approximately 25 × 20 cm. The mass was firm, nontender, and immobile with respect to underlying structures. Pulmonary, cardiovascular, and genitourinary examinations were unremarkable.

Ultrasonography revealed a complex cyst of indeterminate origin measuring 21 × 17.1 cm, right hydronephrosis, a uterine myoma measuring 3.48 × 3.29 cm, and moderate ascites. Computed tomography (CT) identified a homogeneous abdominal mass with a cystic component, presumed to be ovarian in origin, accompanied by mild right hydronephrosis. A complete blood count indicated a white blood cell count of 6.2×10^3 cells/μL, a haemoglobin level of 8.4 g/dL, and a platelet count of 180×10^3/μL. Biochemical analysis revealed the following blood concentrations: urea, 4.46 mmol/L; creatinine, 19.9 μmol/L; sodium, 141 mmol/L; potassium, 3.8 mmol/L; and AST (aspartate aminotransferase), 17.2 U/L.

The initial working diagnosis was Meigs syndrome, and the gynaecologist prepared the patient for exploratory laparotomy with the potential for tumour excision. Intraoperatively, a large abdominal tumour originating from the distal stomach was encountered. The tumour occupied the entire abdominal cavity, causing displacement of the stomach and spleen. Mild ascites was present, but the liver, ovaries, and bowels appeared normal, with no evidence of peritoneal involvement.

The surgical oncology team was consulted and took over the procedure. The operative steps included the discovery of a 22 × 18 cm tumour, weighing 9.6 kg, growing from the distal body of the stomach without invasion into adjacent structures (Figure 1). Approximately 400 mL of straw-coloured ascites was aspirated. A distal gastrectomy, ensuring the remaining gastric stump was supplied by the short gastric vessels, was performed en bloc with tumour excision, accompanied by a total omentectomy and a D2 lymphadenectomy (Figure 2). Intraoperative frozen section analysis for tissue diagnosis was not possible.

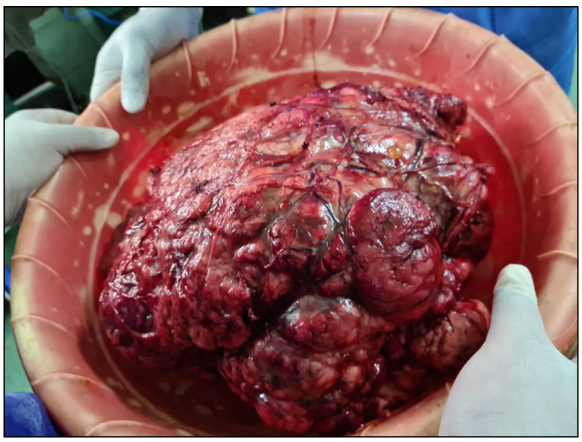

Figure 3. Excised gastric tumour (9.6 kg

The tumour, attached by adhesion bands but not invasive, was removed; however, an iatrogenic injury to the left hemidiaphragm occurred and was subsequently repaired with nylon sutures. An intercostal drainage tube was placed in the left fourth intercostal space. The excised tumour (Figure 3) was sent for histopathologic examination.

Gastrointestinal continuity was reestablished through a Roux-en-Y, antiperistaltic, retrocolic gastrojejunostomy, along with an end-to-side jejunostomy (Figure 2). Finally, the abdominal cavity was lavaged with warm saline, and solitary drains were placed in the gastric bed and pelvis, respectively.

The patient recovered well postoperatively, experiencing no significant complications such as infection or bowel leakage. She was cared for in the intensive care unit for 2 days following the procedure and was discharged from the hospital on the seventh day. The histopathology report indicated that the resection margins were free of tumour cells. The immunohistochemistry report confirmed a diagnosis of gastric GIST, with both CD117 and DOG1 (discovered on GIST 1) markers testing positive. The tumour was classified as high risk, characterized by a mitotic rate >5 per high-power field and a diameter >10 cm.

Postoperative management included a 6-week follow-up visit at the surgical clinic to evaluate for adequate wound healing. Subsequently, the patient was referred to the Cancer Diseases Hospital for treatment with imatinib. She will undergo regular follow-up, consisting of history taking and physical examinations, every 6 months for 2 years, followed by check-ups every 6 to 12 months at the Cancer Diseases Hospital. Additionally, CT scans will be conducted every 3 to 6 months for 3 to 5 years to monitor for any recurrence or progression of the disease.

Discussion

Although the stomach is the most common site for GISTs, gastric GISTs have the poorest prognosis, followed by those in the small intestine.[7] Common sites for gastric GISTs include the body (70%), antrum (15%), and cardia (15%).[7] Our patient had a GIST originating in the body of the stomach. Most GISTs are incidentally discovered during procedures for other indications, such as endoscopic and surgical procedures.[11] Patients with GISTs often do not present with symptoms until complications arise or metastases manifest.[12] Our patient presented with weight loss and early satiety and was initially considered for a laparotomy to potentially excise an abdominal tumour suspected of causing Meigs syndrome. Our patient, at 55 years of age, was younger than the established median age range at GIST diagnosis (60-65 years).[8]

GIST diagnosis requires comprehensive history-taking and physical examination, complemented by imaging to evaluate the resectability of the primary tumour and the existence of unresectable locally advanced or metastatic disease.[13] The detection of a homogeneous abdominal mass should have prompted suspicion of a GIST in our patient. The histopathologic diagnosis of a GIST is established by demonstrating the presence of CD117 or PDGFA via immunohistochemical analysis.[4] However, a preoperative histopathologic diagnosis is essential if the tumour is inoperable and requires neoadjuvant therapy or palliative adjuvant therapy with tyrosine kinase inhibitors, such as imatinib.[14] The immunohistochemistry results for our patient indicated DOG1 (used to identify CD117-negative GISTs) and CD117 positivity in a high-risk tumour with >5 mitoses per high-power field and a diameter >10 cm; yet, this tumour was operable.[15],[16] Given the operability of our patient's abdominal mass, surgery was the initial treatment offered, to be supplemented with adjuvant imatinib therapy, given the associated high-risk factors.

Beham et al.[16] found that patients with gastric GISTs who underwent surgery with negative margins but did not receive adjuvant tyrosine kinase inhibitor treatment had a 5-year survival rate of 35%, accompanied by a tumour recurrence rate of 66%. Our patient was referred for adjuvant therapy at Zambia's national Cancer Diseases Hospital. She will undergo follow-up involving history-taking and physical examinations every 6 months for 2 years, then every 6 to 12 months thereafter.[17] Additionally, CT will be performed every 3 to 6 months for 3 to 5 years.[17]

Conclusions

The stomach is the most common site of origin for GISTs, which are often incidentally discovered during surgery or based on imaging findings. Surgical resection with negative margins remains the preferred treatment. High-risk GISTs that are incidentally detected intraoperatively require adjuvant therapy with tyrosine kinase inhibitors following complete surgical resection.

References

1. Bischof DA, Kim Y, Blazer DG 3rd, et al. Surgical management of advanced gastrointestinal stromal tumors: an international multi-institutional analysis of 158 patients. *J Am Coll Surg*. 2014;219(3):439-449. doi:10.1016/j.jamcollsurg.2014.02.037 [View Article] [PubMed]
2. Boşoteanu M, Boşoteanu C, Deacu M, Aşchie M. Differential diagnosis of a gastric stromal tumor: case report and literature review. *Rom J Morphol Embryol*. 2011;52(4):1361-1368. [PubMed]

3. Stamatakos M, Douzinas E, Stefanaki C, et al. Gastrointestinal stromal tumor. World J Surg Oncol. 2009;7:61. doi:10.1186/1477-7819-7-61 [View Article] [PubMed]
4. Altinas Y, Bayrak M. Gastrointestinal stromal tumor: diagnosis and management. *Int J Surg Res Pract*. 2019;6(1):093. doi:10.23937/2378-3397/1410093 [View Article]
5. Flores-Funes D, Lirón-Ruiz RJ, Pérez-Guarinos CV, et al. Clinical and pathological features of gastrointestinal stromal tumors (GIST) in a single institution: a descriptive study and review of the literature. *Cir Esp (Engl Ed)*. 2017;95(7):391-396. doi:10.1016/j.ciresp.2017.06.005 [View Article] [PubMed]
6. Rodrigues JBSR, Campanati RG, Nolasco F, Bernardes AM, Sanches SRA, Savassi-Rocha PR. Pre-operative gastric gist downsizing: the importance of neoadjuvant therapy. *Arq Bras Cir Dig*. 2019;32(1):e1427. doi:10.1590/0102-672020180001e1427 [View Article] [PubMed]
7. Popivanov G, Tabakov M, Mantese G, et al. Surgical treatment of gastrointestinal stromal tumors of the duodenum: a literature review. *Transl Gastroenterol Hepatol*. 2018;3:71. doi:10.21037/tgh.2018.09.04 [View Article] [PubMed]
8. Kale SS, Sachdev MS, Ismail MK, Davila R, Tombazzi CR. A case and literature review of complicated gastrointestinal stromal tumors. *Gastroenterol Hepatol (N Y)*. 2008;4(9):650-657. [View Article] [PubMed]
9. Đokić M, Novak J, Petrič M, Ranković B, Štabuc M, Trotovšek B. Case report and literature review: patient with gastroduodenal intussusception due to the gastrointestinal stromal tumor of the lesser curvature of the gastric body. *BMC Surg*. 2019;19(1):158. doi:10.1186/s12893-019-0608-3 [View Article] [PubMed]
10. Shah K, Chan KKW, Ko YJ. A systematic review and network meta-analysis of post-imatinib therapy in advanced gastrointestinal stromal tumour. *Curr Oncol*. 2017;24(6):e531-e539. doi:10.3747/co.24.3463 [View Article] [PubMed]]
11. Madhavan A, Phillips AW, Donohoe CL, et al. Surgical Management of gastric gastrointestinal stromal tumours: comparison of outcomes for local and radical resection. *Gastroenterol Res Pract*. 2018;2018:2140253. doi:10.1155/2018/2140253 [View Article] [PubMed]
12. El-Menyar A, Mekkodathil A, Al-Thani H. Diagnosis and management of gastrointestinal stromal tumors: an up-to-date literature review. *J Cancer Res Ther*. 2017;13(6):889-900. doi:10.4103/0973-1482.177499 [View Article] [PubMed]
13. Marcella C, Shi RH, Sarwar S. Clinical Overview of GIST and its latest management by endoscopic resection in upper GI: a literature review. *Gastroenterol Res Pract*. 2018;2018:6864256. doi:10.1155/2018/6864256 [View Article] [PubMed]
14. Lamba G, Gupta R, Lee B, Ambrale S, Liu D. Current management and prognostic features for gastrointestinal stromal tumor (GIST). *Exp Hematol Oncol*. 2012;1(1):14. doi:10.1186/2162-3619-1-14 [View Article] [PubMed]
15. Ahmed M. Recent advances in the management of gastrointestinal stromal tumor. *World J Clin Cases*. 2020;8(15):3142-3155. doi:10.12998/wjcc.v8.i15.3142 [View Article] [PubMed]
16. Beham AW, Schaefer IM, Schüler P, Cameron S, Ghadimi BM. Gastrointestinal stromal tumors. *Int J Colorectal Dis*. 2012;27(6):689-700. doi:10.1007/s00384-011-1353-y [View Article] [PubMed]
17. Koo DH, Ryu MH, Kim KM, et al. Asian consensus guidelines for the diagnosis and management of gastrointestinal stromal tumor. *Cancer Res Treat*. 2016;48(4):1155-1166. doi:10.4143/crt.2016.187 [View Article] [PubMed]

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