CASE REPORT

Primary greater omental GIST presenting with acute intra-abdominal haemorrhage

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SUMMARY

Primary omental gastrointestinal stromal tumours (GISTs) are classified as extra-GISTs with a reported incidence of <1% of all GISTs. There are only a few cases in the English literature reporting omental GIST with none reporting acute intra-abdominal haemorrhage as a presenting feature. We report a case of a 67-year-old man who presented with haemorrhage from a primary omental GIST requiring emergency laparotomy.

BACKGROUND

Gastrointestinal stromal tumours (GISTs) account for <1% of all gastrointestinal tumours. However, they are the most common sarcomatous tumours of the GI tract. They can appear anywhere along the GI tract with the most common location being the stomach (40%–60%), small intestine (30%) and colon (15%). Rarely, GISTs can occur in extraintestinal locations such as the mesentery, omentum and retroperitoneum. Tumours arising from the omentum are exceedingly rare with a reported incidence of <1%. The presentation of GISTs can be highly variable and non-specific with the most common presentation of an omental GIST being a palpable abdominal mass. To our knowledge, this is the second reported case of a primary omental GIST presenting with acute haemorrhage. The rarity of this condition mandates it to be brought to the attention of practising surgeons and adds to the small pool of data regarding extra-GISTs (EGISTs) solidifying its educational importance.

CASE PRESENTATION

A 67-year-old man who is known to have diabetes, hypertension and ischaemic heart disease presented to our institution with a 4-day history of gradual onset, vague, upper abdominal pain that progressively became generalised. This pain was associated with constipation for 1 day with no flatus. No other associated symptoms such as nausea, vomiting, abdominal distension, anorexia or fever were elicited. His drug history was significant for insulin R and enalapril for which he has no compliant. He was not taking any beta blockers or anticoagulants, such as aspirin.

Examination demonstrated an elderly male in no obvious painful distress. His vital signs were all within normal limits. Abdominal examination revealed an obese but soft, non-tender abdomen without guarding or rebound tenderness associated with hypotympanic bowel sounds. The

remaining physical examination including digital rectal examination were unremarkable. Initial full blood count revealed a normocytic anaemia with an haemoglobin (Hb) of 8.6, mean corpuscular volume (MCV) of 90. He then went on to have a CT abdomen which demonstrated a 10 cm×9 cm mass attached to or arising from the greater curvature of the stomach with free fluid noted in the abdomen, features suggestive of a gastric GIST. On the basis of the CT findings and the haemodynamic stability of our patient, he then underwent an upper endoscopy which revealed a mild erosive reflux oesophagitis with no evidence of a gastric GIST.

While being monitored on the surgical floor, he became tachycardic, his repeat Hb dropped to 7 and review of the CT suggested that the free fluid was most likely blood. Based on these factors, our patient was taken for an emergency laparotomy.

INVESTIGATIONS

An initial full blood count revealed a Hb level of 8.6 g/dL, with an MCV of 90. His white blood cell and platelet counts were within normal limits as were his renal function tests.

A CT scan of his chest abdomen and pelvis was performed which demonstrated a 10.2 cm×9.0 cm mass arising from or attached to the greater curvature of the stomach with free fluid noted in the abdomen (figures 1 and 2). There was no evidence of any other masses or signs of metastatic disease.

An upper GI endoscopy was performed, revealing a mild erosive oesophagitis with no evidence of a gastric GIST (figures 3 and 4). During observation on the surgical floor, our patient became tachycardic with increasing abdominal pain. A repeat full blood count revealed a haemoglobin of 7 g/dL.

DIFFERENTIAL DIAGNOSIS

1. omental GIST
2. gastric GIST
3. leiomyosarcoma
4. leiomyoma.

TREATMENT

The patient was taken for an emergency laparotomy. Intraoperative findings included 2500 mL of blood noted in the abdomen with bleeding coming from a mass densely adherent to the greater curvature of the stomach. The mass was carefully dissected off the left lobe of the liver and a wedge resection of the stomach along with resection of the

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Figure 1 Axial view showing a 10 cm×9 cm mass arising from or attached to the greater curvature of the stomach.

greater omentum was performed to ensure clear surgical margins (figures 5–7).

OUTCOME AND FOLLOW-UP

Our patient had an uneventful postoperative recovery and was discharged on day 6 of admission. Histology demonstrated a GIST with no involvement of gastric tissue and arising from the omental tissue. The maximum tumour size was 13 cm with necrosis present. Mitoses were $<5/50$ HPF (1/50 HPF), and the surgical margins were negative for tumour, up to 2 cm. Immunohistochemical staining was done and demonstrated the following profile: CD-117 (c-kit) and CD-34 positive, S-100 and smooth-muscle actin negative (figures 8 and 9). He was started on

Figure 2 Sagittal view demonstrating a large mass closely adherent to the greater curvature of the stomach with free fluid in the abdomen.

Figure 3 Upper GI endoscopy demonstrating no evidence of a gastric gastrointestinal stromal tumours.

imatinib therapy, and at a 1-year follow-up, he is completely asymptomatic with no evidence of recurrence or metastatic disease.

Figure 4 Haemorrhagic free fluid from ruptured primary omental gastrointestinal stromal tumours.

Figure 5 Mass being carefully dissected off left lobe of liver.

DISCUSSION

GISTs comprise <1% of all tumours of the GI tract. This being said, they are the most common, approximately 80%, mesenchymal tumour of the GI tract.^{1,2} They most frequently occur in the stomach (40%–60%) followed by the small intestine (30%) and the colon (15%) and <1% in the oesophagus and appendix.³

Figure 6 Mass excised along with greater omentum and wedge resection of stomach (artery forceps).

Figure 7 Cut specimen showing haemorrhage inside mass.

Rarely, GISTs originate outside the GI tract and these are termed EGISTs. GISTs arising from the omentum are exceedingly rare with a reported incidence of <1%.⁴

Most EGISTs tend to be clinically silent. However, their presentation tends to be very non-specific with the most common symptom being an abdominal mass.⁵ To our knowledge, this is the second case of an omental GIST presenting with acute intra-abdominal haemorrhage in the English literature.⁶ Omental GISTs in close proximity to the stomach are often diagnosed on radiological imaging as a gastric GIST, as in this case. Approximately, 50% of all omental GISTs are misdiagnosed as extramucosal tumours of the stomach.⁷ Similarly, multiple omental GISTs, due to its rarity, are often diagnosed as multiple small bowel GISTs.⁶ With respect to our case, considering our

Figure 8 Histology showing spindle cell type with no evidence of involvement of gastric wall.

Figure 9 Immunohistochemical staining showing CD-117 and CD-34 positive.

patient's haemodynamic stability, we proceeded to perform an upper GI endoscopy which did not demonstrate any evidence of a gastric GIST. Thus, an omental GIST was suspected. One study suggested that most EGISTs represent GISTs with excessive extramural outgrowth resulting in loss of contact to the external muscle coat of the gut or metastasis from an inoperable GIST.⁸ After careful retrospective review of our specimen, there was no

residual intestinal muscle tissue noted; hence, the diagnosis of a primary omental GIST was made.

The mainstay of treatment for non-metastatic GIST or EGIST is resection as was necessary in this case. Omental GISTs are morphologically and immunohistochemically identical to GISTs arising from the stomach or small intestine. They are tumours comprising either spindle or epithelioid cells usually positive for c-kit (CD-117) and CD-34. They may be positive for smooth muscle actin (SMA) but usually negative for desmin and S-100 protein. As demonstrated in our case, the omental GIST was positive for c-kit and CD-34 and negative for both SMA and S-100.

Due to the rarity of omental GISTs, no specific treatment guidelines exist. Therefore, the management used is that for classical GISTs. Hence, adjuvant therapy with imatinib is reasonable for patients with EGISTs post resection. Unfortunately, there is insufficient data on greater omental GISTs to outline a prognosis and survival for these patients.

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Patient's perspective

I was patiently waiting for the elective surgery date of my abdominal cyst. While waiting, I started to have sudden abdominal pain and admitted into the hospital. I was very scared when I was told by my doctor that the cyst in my tummy has ruptured inside. However, I am very grateful to my team of doctors for their immediate intervention and subsequent care.

Learning points

- Gastrointestinal stromal tumour (GIST) arising from greater omentum is an important differential for a gastric GIST.
- Acute haemorrhage from an omental GIST is an important but rare complication.
- Like other GIST, resection is the mainstay of management for non-metastatic omental GISTs.
- Giant abdominal cyst should be resected urgently without any delay because delayed resection can increase the likelihood of acute haemorrhage in the cyst or rupture into the abdomen, which can further increase the morbidity and mortality of the patient.

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