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Gastrointestinal Stromal Tumour presenting with Hemoperitonium: A Case Report.

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Abstract: *Gastrointestinal stromal tumour (GIST) is the most common mesenchymal tumour of the GI tract, with the majority occurring in the stomach and the small intestine. This is a vascular tumour that primarily presents with GI bleeding (50%), a palpable mass (35%, sometimes with features of intestinal obstruction), pain (20%), and rarely as an incidental finding. Hemoperitoneum as a presentation due to GIST rupture is very rare. We here present a very rare case of spontaneously ruptured GIST of the ileum presenting with peritonitis due to hemoperitoneum.*

Keywords: *GIST, spontaneous rupture, hemoperitoneum, CD 117.*

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I. Introduction

Gastrointestinal stromal tumour (GIST) is the most common mesenchymal tumour of the GI tract expressing c-kit (CD 117), which is a highly specific marker differentiating GIST from other mesenchymal tumours, such as leiomyomas[1-3]. Majority of the GIST occur in the stomach (60%-70%) and small intestine (20%-30%)[2]. Around 10%-30% are asymptomatic[3]. Symptomatic GISTs are generally associated with abdominal pain, GI bleeding or a palpable mass but rarely associated with hemoperitoneum[4]. We here present a very rare case of spontaneously ruptured GIST of the ileum presenting with peritonitis due to hemoperitoneum.

II. Case Report

A 52year old gentleman, with no comorbidities presented to our casualty with complaints of pain abdomen and distension of abdomen for 3 days, and decreased urine output for 1 day. There was no associated fever or obstipation or history of alcohol abuse. On examination, Pulse – 120/ min, B.P. – 80/50 mm of Hg, Abdomen was tense and distended, rebound tenderness and rigidity was present and bowel sounds were absent. X-ray Chest and Abdomen revealed few dilated small bowel loops, but no gas under the domes of diaphragm. Patient was resuscitated with i.v. fluids and i.v. antibiotics were started. A provisional diagnosis of perforation peritonitis was made and patient was taken for emergency exploratory laparotomy. Intaoperatively, to our surprise there was around 1 litre of haemorrhagic fluid, there was mass arising from the small bowel around 150 cm from the DJ flexure measuring 13 x 12 x 7 cm on the anti-mesenteric border, wall of the mass was friable and found to be ruptured and adhered to the parietal peritoneum just above the urinary bladder. Its cavity contained blood clots. The mass was excised intoto with the segment of small bowel (Fig.1), the bits of the rupture wall were meticulously removed from the pelvis and end to end anastomosis was fashioned after giving a thorough abdominal lavage. Post-operatively recovery was uneventful. Histopathology, revealed haemorrhage and necrosis within the tumour, spindle shaped tumour cells with acidophilic cytoplasm and enlarged nuclei, mitotic count of 5 per 50 HPF, positive for CD 34, desmin and smooth muscle actin, weakly positive for CD 117 and negative for S 100 all suggestive of GIST. Case was discussed in tumour clinic and in view of tumour rupture, it was labelled as metastatic GIST and patient was asked be on life-long Imatinib mesylate 400 mg daily.

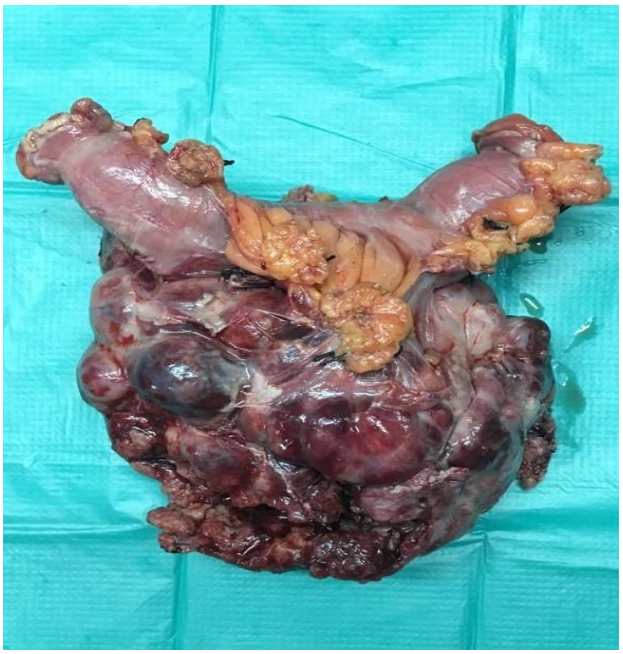

Fig. 1: Showing 13x12x7 cm friable mass with its wall ruptured in the antimesenteric border with the resected segment of small bowel.

III. Discussion

GIST is a vascular tumour that primarily presents with GI bleeding (50%), a palpable mass (35%, sometimes with features of intestinal obstruction), pain (20%), and rarely as an incidental finding[5]. Hemoperitoneum as a presentation due to rupture is very rare. The mechanism underlying hemoperitoneum may be related to bleeding in the tumour leading to hematoma and rupture of the capsule or transudation of blood components from the tumour.

Due to its rarity, diagnosis requires a high index of suspicion. The primary mode of diagnosis and assessment of extent of disease is by contrast enhanced Computed topography (CT) of abdomen and pelvis, revealing a heterogeneously enhancing, exophytic mass in close relation with stomach or bowel wall. MRI is useful in rectal GIST. PET scan is not used to diagnose, but to assess the response to therapy. On Endoscopy, GIST appears as a submucosal mass. Endoscopic-guided fine needle aspiration has been shown to be ~ 80% sensitive in diagnosing GIST. Because GISTs tend to be soft and friable, biopsy carries a high risk of tumour rupture and bleeding[6].

Macroscopically, GIST are usually grey-white in appearance. They are believed to arise from the interstitial cells of Cajal in the muscularis propria and can grow exophytically out into the peritoneum, or endophytically into the lumen of the gut[7]. There are three histological sub-types of GIST. The spindle form is the most common (70%) and consists of uniform intersecting fascicles with eosinophilic cytoplasm. The epithelioid (20%) and the rare mixed type (10%) form shows more rounded cells with nuclear atypia. Approximately 95%of GISTs stain positive for KIT (CD 117) by immunohistochemistry (IHC). Epithelioid GISTs tend to have weaker KIT staining than the spindle cell type. Other markers include CD 34 (70%), smooth muscle actin (30%) and desmin (<5%)[2]. The diagnosis of GIST is based on concordance between morphology and IHC.

Prognostic factors indicating possible malignant potential for GIST include tumour size, mitotic rate and location. Small tumours (<2 cm) with low mitotic rates (<5 per 50 HPF) exhibit benign behaviour, whereas larger tumours (>5cm) with high mitotic rates (>10 per 50 HPF) are associated with malignant behaviour and display higher rates of recurrence after surgical resection. Tumours in the stomach have favourable outcomes relative to small bowel tumours. Of the three factors, mitotic rate is considered the most significant[8]. Tumour rupture before or during surgery also portends a worse outcome manifested by higher rates of peritoneal recurrence and is managed by some surgeons as metastatic GIST.

Surgery remains the only curative option in patients with localised primary GIST. The goal being to achieve negative microscopic margins with an intact tumour pseudocapsule. Wide margins have not been shown to improve outcomes. Lymphadenectomy is not routinely required unless adjacent nodes are obviously enlarged. En bloc resection is needed when adjacent organs are involved[9]. Adjuvant therapy with Imatinib mesylate for 3 years vs 1 year in high risk cases has been found to improve the recurrence free survival and overall survival[10].

IV. Conclusion

Spontaneous rupture is a rare presentation of GIST and preoperative diagnosis is difficult due to its acute presentation, giving less time for radiological investigations. GIST should be considered as one of the differentials in a patient presenting with spontaneous hemoperitoneum.

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