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Perforation Peritonitis by a Gastrointestinal Stromal Tumour of Jejunum: A Rare Presentation

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Running Title: *"Gastrointestinal Stromal Tumour of Jejunum presenting with Perforation Peritonitis"*

Abstract: ***Tumour perforation is accepted as a bad prognostic factor along with larger tumour size and higher mitotic index in gastrointestinal stromal tumours (GISTs). The prognosis is worse in patients with tumour perforation or rupture. We report an unusual case of a 5 cm GIST of the jejunum that presented with spontaneous rupture. A previously healthy 46-year-old male patient presented with sudden abdominal pain from last 3 days. Physical examination revealed features of peritonitis, and computed tomography showed a heterogeneous solid mass measuring approximately 5 cm x 5.5 cm arising from the distal jejunum with mild fluid in the peritoneal cavity. The mass was diagnosed as a GIST originating from the gastrointestinal tract. Emergency laparotomy was performed and intraoperative findings showed mild hemoperitoneum and an outgrowing mass from the distal jejunum. The tumour had ruptured away from the wall of the distal jejunum though no active bleed was note. Surgical resection of the tumour was performed and the patient was discharged home uneventfully on the third postoperative day. The immunohistochemical characteristics of the tumour revealed it to be a GIST.***

Keywords: Gastrointestinal stromal tumour (GIST), peritonitis, exophytic.

1. Introduction

Gastrointestinal stromal tumours (GISTs) are mesenchymal tumours of the gastrointestinal (GI) tract, that originate from Cajal interstitial cells **[1].**GISTs represents 80% of mesenchymal GI tumours and 0.1-3% of all GI malignancies **[2,3].** Gastrointestinal stromal tumours (GISTs) are the most common mesenchymal tumour of the gastrointestinal (GI) tract **[1].** Most GISTs are >5 cm in diameter at the time of diagnosis, with a diameter of 10 cm being associated with a higher risk of local or distant metastasis. Gastrointestinal bleeding is the most common presentation (50%) of GISTs and is usually associated with ulceration of the tumour into the lumen **[4,5].** In addition to the tumour size, mitotic rate and tumour location, tumour rupture is believed to be a prognostic factor for the outcome of patients with a GIST. The clinical symptoms of a GIST range from mild to severe, and complications include vague abdominal pain, hematemesis, and intestinal obstruction. However, overt peritonitis caused by GIST rupture is very uncommon. This paper describes the clinical course of acute abdomen caused by a rare case of a ruptured exophytic small bowel GIST

2. Case Presentation

A 46-year-old man with no known medical or surgical comorbidities presented to the emergency surgery department at Sheri- Kashmir Institute of Medical Sciences at 7:00 am with complaints of pain abdomen from last 3 days. He had an episode of upper GI bleed on 24/12/2019, which was managed conservatively and EGD done that time was showing features of Erosive Gastritis. He had no previous history of any surgery or any chronic disease. He denied any previous medication, nausea, vomiting, changes in bowel habit, melena, or weight loss. His medical and family histories were unremarkable. His body temperature was 36.5°C. His blood pressure was 126/74 mm-Hg and pulse rate was 110 beats/min. Saturation was 93% on room air. Auscultation indicated clear lungs, and cardiac examination demonstrated no murmurs, rubs, gallops, or crunch.

Physical examination revealed Glasgow Coma Score of 15/15, that the abdomen was distended, with diffuse tenderness and without rebound tenderness. Nasogastric tube was secured and aspiration was done Blood analysis revealed a white blood cell count of 11000/µL, initial haemoglobin level of 10.6 g/dL and platelet count of 223000/µL. The laboratory tests for blood glucose 99 mg/dl, electrolytes were sodium 125, potassium 3.4, urea 22, creatinine 0.80, and amylase was 36, and the liver function tests were within normal range.

Ultrasonography **[Fig.1]** revealed an exophytic predominantly hypoechoic lesion with central anechoic area with extensive surrounding soft tissue and mesenteric stranding with no definite vascularity on colour Doppler. The lesion was clearly separate from bowel lumen along the antimesenteric border. Size of the lesion was 4.8x5.0x4.9cm

approximately. No features of intestinal obstruction were present.

The patient was admitted to general surgery in-patient clinic with a pre-diagnosis of a possible lesion of intestinal origin with surrounding inflammatory changes and was advised contrast study. Unenhanced Computed tomography (CT) scan [**Fig.1(a)]** revealed free fluid in cul-de-sac with evidence of mass lesion which seemed to be exophytic in location in distal jejunum with evidence of surrounding soft tissue and fat stranding. An abdominal enhanced CT scan **[Fig.1(b)]** revealed the presence of a heterogeneously enhancing solid mass measuring approximately 5 x 5.5 cm attached to the distal jejunal antimesenteric border and mild fluid in the peritoneal cavity. The mass was radiologically suspected to be a GIST arising from the jejunum and the fluid in the peritoneum indicated possible tumour rupture. We suspected the patient's condition to be peritonitis induced by tumour rupture and decided to perform emergency laparotomy under general anaesthesia for perforation peritonitis induced by an exophytic jejunal lesion with laparotomy and resection anastomosis of jejunum and subsequent peritoneal lavage.

Intra-operative findings

- A midline abdominal incision was given and abdominal layers opened.
- Lot of adhesions were present between omentum and the parietal wall
- About 100ml of blood-tinged purulent fluid was present in peritoneal cavity
- Around 5 x 5cm exophytic mass lesion **[Fig.2] & [Fig.4**] was present in distal jejunum adherent to sigmoid colon with perforation noted at the apex.
- Resection anastomosis of jejunum **[Fig.2]** was done and the specimen was sent for histopathological analysis and immunohistochemistry. The tumour had ruptured **[Fig. 3]** away the wall of the small intestine but active bleeding was not observed. A total of 100 ml of blood with intermixed fluid was evacuated from the abdominal cavity.
- Haematoxylin & Eosin staining [**Fig.5]** showed spindle-shaped cell proliferation and immunohistochemical reports indicated that the tumour was strongly and diffusely positive for CD117, DOG1 and bcl-2 while totally negative for the S-100 protein, smooth muscle actin, and CD34. These findings supported the diagnosis of a GIST. The patient recovered well without postoperative complications and was discharged on the third postoperative day.
- GIST of the jejunum was diagnosed and treated with imatinib mesylate, 400 mg daily, after the 20th postoperative day. The patient signed a written informed consent form granting his permission for publication of his details in this case report

3. Discussion

Gastrointestinal stromal tumours (GISTs) represent the most common mesenchymal tumour of the gastrointestinal tract. GISTs are visceral tumours which originate from interstitial cells of Cajal arising from any site of the GI tract. Around 90% of GISTs occurs in the age group of greater than 40 years old with male dominance. Approximately 60-70% of cases occur in the stomach, 25-35% in the small intestine, and 10% in the jejunum, whereas the oesophagus, colon, rectum, and appendix are rarely affected **[6]**. Most GISTs are clinically silent till they grow large, bleed, rupture, cause mechanical obstruction or act as a lead point for intussusception. One of the most common clinical manifestations for symptomatic GISTs is occult GI bleeding from mucosal ulceration. In most cases, strong positivity is observed for KIT (CD117) (approximately 95%) and CD34 (approximately 70%) **[7].** It is difficult to diagnose a jejunal GIST preoperatively due to the nonspecific and variable clinical symptoms, and it is also difficult to distinguish the tumour based solely on images. GISTs present on CT as exophytic rounded masses with heterogeneous enhancement. GISTs originating in the jejunum seldom cause perforation and acute diffuse peritonitis **[8].** Large GISTs nearly always outgrow their vascular supply, leading to extensive areas of necrosis and haemorrhage **[9].** Surgical resection is the "gold standard" for therapy of GIST. The primary goal of surgery is complete resection of the disease **[10].** Primary tumours are typically exophytic (79%), larger than 5 cm (84%), and heterogeneously enhanced on CT scans (84%). Metastatic seeding mostly occurs to the peritoneal cavity and liver, followed by to the lungs and bones. However, a GIST in the small intestine rarely causes hemoperitoneum. The prognosis is dismal when the tumour presents with symptoms or signs such as perforation or rupture, multifocal location, or metastatic lesions. Bleeding into the peritoneal cavity on account of a ruptured GIST can lead to acute abdominal pain presenting as a surgical emergency **[10].** 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. In the present case, bleeding in the tumour leading to rupture of the capsule may have caused hemoperitoneum. Because of their high vascularity, GISTs are frequently associated with GI bleeding and have been associated with severe GI haemorrhage, requiring either embolization or emergency surgery. Our case study suggests that a GIST should be considered when acute non-traumatic hemoperitoneum is present, particularly if CT detects a heterogeneous mass with high vascularity.

4. Conclusion

Spontaneous rupture is a rare presentation of a GIST and preoperative diagnosis is difficult because of the absence of pathognomonic signs or symptoms. A GIST should be suspected whenever there is presentation of sudden abdominal pain in patients with an intra-abdominal mass. We reported the case of a man with a GIST originating in the jejunum that presented as spontaneous rupture and led to hemoperitoneum.

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Statement of Ethics
This case is exempt form ethics committee approval because this is a retrospective analysis of a single clinical case and does not meet the criteria for research.

Conflict of interest statement:
The authors declare that they have no conflicts of interest.

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Author contributions:
Dr Musadiq: conceptualization, acquisition of data & writing original draft presentation. **Dr Mudassir & Dr Omar:** Both authors were the operating surgeons for the present case.All the surgical aspects of the case from preoperative, intraoperative & postoperative management were dealt by them. **Dr Aliya:** Pathological & immunohistochemical aspects of the case.

Data Availability Statement:
All data underlying the results are available as part of the article. Further enquiries can be directed to the corresponding author.

Consent:
Proper consent of the patient was taken for publication of the images and required data in any medical journal.

Legends:
Fig.1. Grey scale ultrasound showing a large hypoechoic mass lesion with anechoic (necrotic) centre & surrounding inflammatory stranding, in close relation to small bowel mesentery.
Fig.1(a). Unenhanced CT showing predominantly exophytic mass lesion arising in relation to antimesenteric surface of jejunum (solid arrows).
Fig.1(b). Enhanced CT showing heterogeneously enhancing predominantly exophytic mass arising from jejunum (solid arrows) with surrounding inflammatory changes.
Fig.2. Intraoperative image showing exophytic mass lesion (solid arrow) attached to the antimesenteric surface of jejunum.
Fig.3 & Fig.4. Resected specimen of jejunum showing the sites of perforation (solid arrows).
Fig.5. Histopathological section of the tumour showing predominant spindle cells.

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