ATYPICAL PRESENTATION OF GASTROINTESTINAL STROMAL TUMOURS WITH PNEUMOPERITONEUM: A CASE REPORT

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ABSTRACT

Gastrointestinal stromal tumours (GISTs) are benign mesenchymal tumours of the gastrointestinal tract (GIT). They originate from interstitial Cajal cells on intestinal pacemaker cells that arise from the muscularis propria of the gastrointestinal tract wall. GISTs are characterized by the expression of c-KIT protein (CD 117, stem cell factor receptor). c-KIT protein, detected via immunohistochemical analysis is the primary diagnostic criterion for a GIST. The stomach is the most frequent site of GIST occurrence. The rupture of GIST of the peritoneal cavity is a critical complication.

KEYWORDS

Jejunum, GIST, Perforative Peritonitis.

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INTRODUCTION

Gastrointestinal stromal tumours are mesenchymal tumours of the digestive tract that originate from intestinal Cajal cells and account for 0.1-3% of all gastrointestinal tumours. They are usually located in the stomach and small intestine, but they can be located anywhere in the gastrointestinal tract, including the omentum and peritoneum. GISTs are characterized by the expression of c-KIT protein (CD 117, stem cell factor receptor.¹) Generally, GISTs have a silent behaviour and are diagnosed incidentally. Approximately 40% of GIST cases cause intestinal bleeding.² Perforation is rarely observed in GISTs; however, we present a case of perforated GIST located in the jejunum as a rare cause of acute abdomen.

CASE REPORT

A 52-year-old male presented in the emergency department with acute abdominal pain during previous 20h. The patient had no complaint of nausea or vomiting; however, the patient did have a history of diabetes mellitus type 2.

On physical examination, patient was conscious and of normal built. Pallor, cyanosis, icterus and oedema were absent. He was normotensive (124/70 mmHg), had tachycardia (110/min), fever (102.4°F) and hurried respiration (25/min). Abdominal examination revealed distension, board like rigidity, marked rebound tenderness, absent liver dullness and inaudible bowel sounds. Hernia sites were normal. Per rectal examination did not reveal any significant abnormality.

Examinations of other systems were within normal limits. A provisional diagnosis of hollow viscus perforation was made.

Exploratory laparotomy was planned. Haematological examination revealed mild anaemic with neutrophilic leucocytosis [Haemoglobin – 9.8 g/dl, Total count- 14,000/cu.mm (N85, L11, E10, B0, M0)]. Blood sugar (113 g/dl), liver function tests and serum electrolytes (Na-136 meq/lit, K- 4.2 meq/lit) were within normal limits. Viral markers were non-reactive. Abdominal roentgenogram showed free gas under both domes of diaphragm with diffuse ground glass opacity. Excessive gas in the abdomen with free fluid was noted in abdominal sonography. The patient was resuscitated with intravenous fluids, Ryles tube and antibiotics. Following adequate resuscitation, the patient was put up for operation. Midline laparotomy revealed purulent free fluid with flakes.

On aspiration and removal of the flakes and fluid, a purplish coloured firm growth with everted margins (Fig. 1), measuring 5*4 cm was found in the anti-mesenteric border of the jejunum, fifty cm from the duodeno-jejunal flexure (Fig. 2). The growth had a central perforation with intestinal contents effusing through the rent). All other organs were found to be normal. The growth was resected with five cm margin (Fig. 3) and an end to end, double layer anastomosis was performed (Fig. 4, 5). Thorough peritoneal lavage was done with warm normal saline and abdomen was closed in layers. A tube drain was placed in the hepatorenal pouch of Morrison.

The specimen was sent for histopathological examination. Post-operative period was uneventful and the patient was discharged on the tenth post-op day after suture removal and he was referred to the department of oncology for adjuvant chemotherapy. Gross examination of the specimen revealed an outward bulging mass, which was centrally necrotic and contained haemorrhagic-necrotic material (Fig. 6) Histopathological assessment of the tumour showed a well delineated epithelioid gastrointestinal stromal tumour originating from the small bowel wall. Tumour cells did not infiltrate the mucosa but only muscularis propria. Based on the size, location and the number of mitoses (<5/50 HPFs), it was considered as a low malignant potential.

There was no lymph node metastasis. It confirmed complete resection with clear surgical margins.

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INTRA OPERATIVE IMAGES

Fig. 1: Bowels Clamps in Situ after Mobilising the Segment of Jejunum to be Resected

Fig. 2: Firm Growth with Everted Margins found in the Anti-Mesenteric Border of Jejunum

Fig. 3: Jejunum after Resection of the Tumour

Fig. 4: Double Layer Anastomosis of the Resected Segment of Jejunum

Fig. 5: After Anastomosis

Fig. 6: Resected Specimen of Jejunum Containing the Tumour

IMAGING

X-Ray abdomen erect shows air under diaphragm

Fig. 7: Erect X-ray Abdomen showing Air Under Diaphragm

DISCUSSION

GIST is a benign mesenchymal tumor arising from the intestinal cells of Cajal. About 60–70 % of the GIST arises from the stomach, 20–30 % from the small intestine, 10% from the rest of the GIT. The incidence of the GIST being 1.5/1,000,000/year, there is a slight male preponderance. Most of the GISTs are submucosal lesions and 15 % may be exogastric as in our case, and less than 5 % can be both extra- and intraluminal (Dumbbell). GIST can also be multicentric in less than 5 % of the cases. Small GISTs are often asymptomatic and are detected incidentally. They become symptomatic because of their large size and their tendency to ulcerate and bleed³. Most common presenting signs and symptoms include abdominal pain, GI bleeding, and palpable mass. Very rarely a pedunculated GIST can cause obstruction to the pylorus and act as a lead point to intussusception.

Histologically GIST can be benign (90 %) or malignant (10 %). GISTs with 1–5 mitosis/ 50 HPF are classified as a low grade, and GISTs with a mitosis of 10/10 HPF are classified as high-grade group. The negative prognostic factors include size greater than 5 cm, high mitotic index and a positive resection margins. Other poor prognostic factors include tumor rupture, more distal location, high cellularity, tumor necrosis, and presence of metastasis⁴. The histological factors with a favourable prognosis include low mitotic index, size less than 5 cm and lack of infiltration of the adjacent organs. Immunohistologically most GISTs have CD 34 antigen and CD 117 antigen. CD 34 protein being a hematopoietic progenitor cell occurs in a variety of mesenchymal tumours.

In the gastrointestinal tract, CD 117-positive cells (Interstitial Cajal cells) are autonomic nerve-related gastrointestinal pacemaker cells that regulate intestinal motility. Because of the immunohistochemical and ultrastructural similarities of Cajal cells and GISTs, a histogenetic origin of GISTs from Cajal cells has been proposed. A few previous studies have investigated ruptured GISTs by means of several radiologic modalities such as

ultrasonography and computed tomography. A literature search yielded only 9 reports of ruptured primary GISTs to date.⁴ Cegarra-Navarro et al reported 5 patients with a ruptured primary GIST and 1 patient with a ruptured metastatic GIST.⁵ We did not evaluate ruptured metastatic GISTs. One of the nine patients' tumours was small bowel location and another one was transverse mesocolon location; the others were gastric location.

Seven of the nine patients' ruptured GISTs were spontaneous; the others were result of the trauma. The size of 3 of the 9 previously reported GISTs was less than 10 cm and the 6 remaining GISTs were larger than 10 cm. As in most of the other cases reported, the mass found in our patient was larger than 10 cm. seven of the previously reported GISTs exhibited low mitotic activity (less than 5 mitoses per 50 HPF). Two of the reported ruptured primary GISTs demonstrated a high mitotic index (20 mitoses per 50 HPF). As in most of the other cases reported, our patient's tumour was characterized by borderline mitotic activity. All primary GISTs that were previously reported except 2 demonstrated low or borderline mitotic activity, but all had a large cystic and necrotic component.⁶ We may speculate that the rupture of primary GISTs involves the cystic and necrotic tumour components but not high-grade mitotic activity.

Most of the patients with a ruptured GIST presented for urgent care and all patients underwent emergency laparotomy. Segmental small bowel or gastric resection is standard treatment for perforated local GIST. Imatinib is being evaluated as adjuvant treatment following surgery, especially in high malignant potential cases.⁷ To our knowledge, a ruptured GIST with pneumoperitoneum has not been previously reported. We describe surgical correction, and histopathologic findings of a ruptured primary GIST⁸. In our patient, the operation was successful, and he did well after surgery.

CONCLUSION

Emergency presentation with a GIST is not uncommon and one of its manifestations is acute abdomen secondary to intraperitoneal rupture or perforation of a primary GIST of the small intestine. Emergency surgery is mandatory and should achieve radical resection.

Primary anastomosis has been reported to be safe. Since there is an increased risk of recurrence after spontaneous intraperitoneal rupture/perforation of GISTs, patients should be evaluated by a multidisciplinary team in order to assess the indications for imatinib adjuvant therapy and for close monitoring and follow-up.

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