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A Case of Perforated Jejunal GIST

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Abstract: We report a case of a seventy-year-old gentleman presenting with pain abdomen for 3 days which was sudden in onset, non-radiating, colicky in nature and gradually progressing to diffuse abdominal pain. Emergency chest x-ray (PA view) showed "Gas under Diaphragm" signifying suspicion of intestinal perforation. Exploratory laparotomy showed a thickened segment of jejunum 20 cm from DJ flexure with a perforation at antimesenteric border having a diameter of 2 cm which was resected with 5 cm margin and anastomosed. Histopathological and immunohistochemical examination diagnosed it to be a CD117 (KIT) positive Jejunal (Small Intestinal) Epithelioid GIST with high mitotic count and atypia ($pT_3N_0M_x$), with negative resection margins and reactive lymph node hyperplasia. The diagnosis along with its site of presentation and certain peculiarities constitute a rarity.

Keywords: Gastrointestinal Stromal Tumour, Intestinal Perforation**,** Jejunal GIST.

INTRODUCTION

Gastrointestinal stromal tumours (GISTs) are a type of mesenchymal tumour of the gastrointestinal (GI) tract [1]. These mesenchymal tumours were initially thought to be extremely rare, but as pathologic techniques and awareness have improved, clinicians are reporting GISTs at a higher incidence. GISTs may be found throughout the gastrointestinal tract, from the oesophagus to the internal anal sphincter.

The most common gastrointestinal location is the stomach (50% to 60%), followed by the small intestine (20% to 25%), rectum (5%), and oesophagus (2%). Other less common locations where GIST has been identified include the omentum and genitourinary tract [2-5]. GIST originates from the interstitial cells of Cajal (ICC). The ICC were identified in 1893 by *Santiago Ramon y Cajal*, who aptly named them "interstitial" cells given their interposition between nerve endings and smooth muscle cells in the gut wall.

CD117 (a tyrosine kinase receptor known as KIT) immunohistochemistry staining is now widely accepted as a criterion for a pathologic diagnosis of GIST [2]. In KIT-negative tumours, the diagnosis continues to rely on histologic features. GISTs contain a variable combination of spindle cells and epithelioid cells. Rarely, GISTs may have myxoid stroma, neuroendocrine features, a signet ring variant, or marked lymphocytic infiltrate. Additional research has identified a new immunohistochemical stain called DOG1—literally *d*iscovered *o*n *G*IST. *Liegl et al* in 2009 examined the sensitivity and specificity of a specific monoclonal antibody, DOG 1.1, directed against DOG1 and suggested that this new antibody may have improved sensitivity compared with KIT for the diagnosis of GIST [6]. Approximately 5% to 15% of GISTs have been found to be negative for KIT mutations and are considered wild-type GIST. Platelet-derived growth factor receptor alpha (PDGFRA) was identified as an alternative oncogene responsible for activation of the intracellular phosphorylation cascade [7].

All GISTs have some ability to metastasize and should not be considered truly benign. Small, incidentally discovered lesions have lower risk for metastasis, but certainly can become invasive if left untreated and the patient is lost to follow-up. A 2002 study by *Fletcher et al* characterized the malignant potential of GISTs, which established primary tumor criteria for predicting disease recurrence, and is widely cited [8]. The mainstay of therapy remains surgical resection. Interestingly, the presence of microscopically positive margins (after macroscopic total resection) has not been demonstrated to confer a worse prognosis in either progression-free survival or overall survival rate [9]. If a pathologic specimen is found to have microscopically positive margins, it does not require additional surgery to obtain R0 resection status. Neoadjuvant as well as adjuvant therapy is based on imatinib, with sunitinib as second-line therapy.

CASE REPORT

A Seventy-year-old gentleman presented to our emergency department with pain abdomen for 3 days. The pain was sudden in onset following breakfast, non-radiating, colicky in nature and gradually progressed to diffuse abdominal pain. No relieving factors were there as the pain was at its peak from the beginning. It was not associated with any nausea, fever, loose stool or prior constipation. Only one episode of vomiting happened after lunch.

There was history of mucoid/slimy discharge with occasional fresh bleeding per-rectum every 2-3 months for last 1 year. He also noticed noticeable weight loss with generalized weakness in last 6 months. Emergency chest x-ray (PA view) showed "Gas under Diaphragm" signifying suspicion of intestinal perforation. He was prepped for operation and vitals stabilized with iv fluids and other ancillary conservative management. Emergency blood biochemistry was not deranged.

Exploratory laparotomy was performed. Intraabdominal pus mixed collection removed. A thickened segment found 20 cm from DJ flexure with a perforation at antimesenteric border having a diameter of 2 cm. The segment of jejunum is resected with 5 cm margin and anastomosis done in two layers. Postoperative period was uneventful. Histopathological examination diagnosed it to be Jejunal (Small Intestinal) Epithelioid GIST with high mitotic count and atypia ($pT_3N_0M_x$). IHC for CD117 came out positive. Resection margins were free and lymph nodes showed reactive hyperplasia.

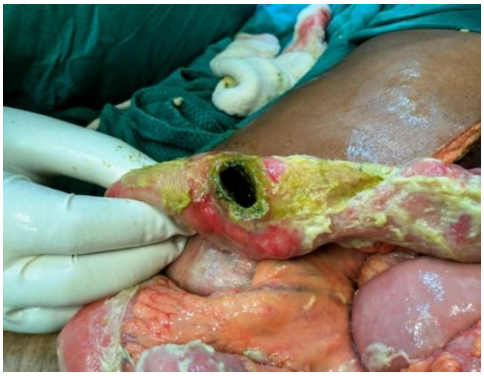

Fig-1: Thickened segment of jejunum showing a perforation (approx. 2 cm diameter)

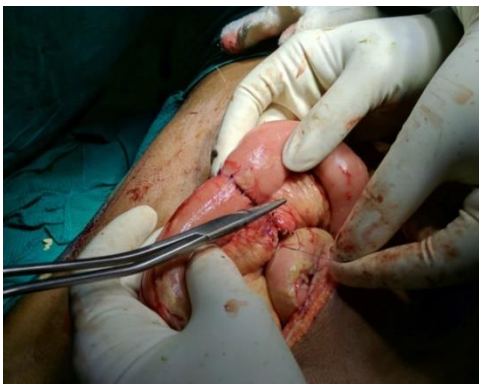

Fig-2: Resected segment of jejunum being anastomosed in two layers

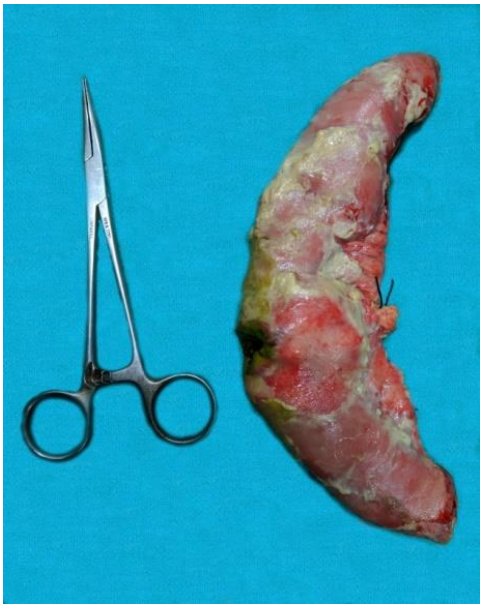

Fig-3: Resected specimen of perforated jejunal GIST

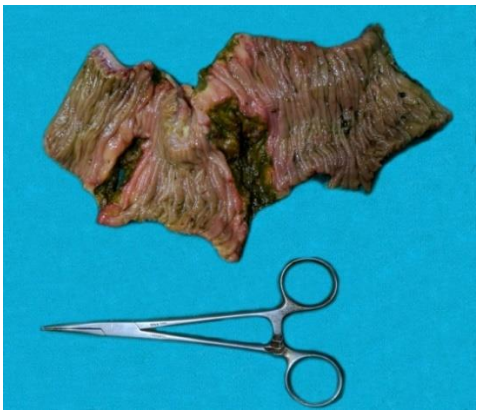

Fig-4: Resected specimen of perforated jejunal GIST (Cut Open)

DISCUSSIONS

The chief objective of outlining this case report was to highlight the rarity of a case. In addition, there are few peculiarities in this case which attracts attention. Over 90% of GISTs occur in adults over 40 years old. The incidence peak of diagnosis is 60–65 years. There is a slight male predominance. Our patient was seventy-year-old.

Gastrointestinal stromal tumours (GISTs) are rare. GISTs comprise only 0.2% of gastrointestinal (GI) tumours and are mainly gastric tumours (50% to 60%). Only 20% of GISTs, i.e. 0.04% of all GI tumours, are small intestinal GISTs and jejunal GISTs are the rarest subtype. Only 10–30% progress to malignancy [10]. Here it was a jejunal GIST.

Jejunal GISTs are typically asymptomatic while small and may be diagnosed incidentally from CT, endoscopy, during surgery or from symptomatic liver metastases. Enlargement causes variable symptomatology; GI bleeding or non-specific GI symptoms such as bloating or early satiety. Around 40% are associated with ulceration, and 28% presenting with overt GI bleeding. Bleeding may be acute (hematemesis or melena) or chronic (anaemia). Around 20% grow large enough to present with pain, a palpable mass or obstruction secondary to intussusception [11]. Here, there was history of occasional (3 – 4 episodes) bleeding per-rectum followed by colicky pain and intestinal perforation which is not common.

The morphology of jejunal GISTs is varied: tumours may be composed of spindle cells (70%), epithelioid cells (20%) or mixed spindle and epithelioid cells (10%). It can also be seen with leiomyosarcomas. Here it was an epithelioid variety of GIST with high mitotic count and atypia which is rare. Finally, a clear resection margin was achieved and the patient made a remarkable recovery.

CONCLUSION

This case presented with sudden, non-radiating, colicky pain abdomen for 3 days. Emergency chest x-ray (PA view) showed “Gas under Diaphragm. Exploratory laparotomy showed a thickened segment of

jejunum 20 cm from DJ flexure with a perforation at antimesenteric border having a diameter of 2 cm which was resected and anastomosed. Postoperative period was uneventful with a good recovery. Histopathological and immunohistochemical examination diagnosed it to be a CD117 (KIT) positive Jejunal (Small Intestinal) Epithelioid GIST with high mitotic count and atypia (pT3N0Mx).

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