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Research Paper

Jejunal GIST presenting as perforativeperitonitis – a case report and review of literature

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ABSTRACT
Gastrointestinal stromal tumours (GIST) are highly frequent mesenchymal tumours of the digestive tract, which mainly affect the stomach and small intestine. GISTs frequently exist with unclear symptoms. Their initial clinical presentation as acute abdomen due to their perforation is particularly rare. We report a case of a 36-year-old male presenting with acute abdomen. The final diagnosis revealed a perforated GIST in the jejunum. In this paper, we report the clinical manifestation as well as computed tomography and histopathological findings helpful for the accurate diagnosis of this rare complication of GIST. Emergency laparotomy and resection of tumour are essential. Following surgical resection, adjuvant tyrosine kinase inhibitor therapy should be considered for prevention of early recurrence.

KEYWORDS:*Gastrointestinal stromal tumour, perforative peritonitis, spontaneous GIST perforation, jejunal GIST*

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I. INTRODUCTION

Gastrointestinal stromal tumours (GIST) are mesenchymaltumours of the digestive tract that originate from interstitial Cajal cells and account for 0.1-3% of all gastrointestinal tumours. They are usually located in the stomach and small intestine [1], but they can be located anywhere in the gastrointestinal tract, including the omentum and peritoneum. Approximately 40% of GIST cases cause intestinal bleeding [2]. It occurs morecommonly in men with a median age of 50–70. GISTs are thoughtto be the result of mutations of proto-oncogene which encodes thecell surface tyrosine kinase receptor. Perforation is rarely observed in GISTs. We present a case of perforated GIST located in the jejunum as a rare cause of acute abdomen.

II. CASE PRESENTATION

A 36 year old male patient presented in the emergency with gradually increasing pain abdomen and vomiting for last four days. He did not have any prior surgical history and no known co-morbidity. There was no history of alcoholism or any other addiction either. Vitals were normal. On clinical examination there was generalised tenderness over the abdomen with rigidity and rebound tenderness. Bowel sounds were absent.

A contrast CT Scan whole abdomen showed free air density in peritoneal cavity suggestive of pneumoperitoneum. Multiple irregular pockets of peritoneal collection with internal air density, hyper-density (hu-45 to 50) and enhancing walls -suggestive of peritonitis.

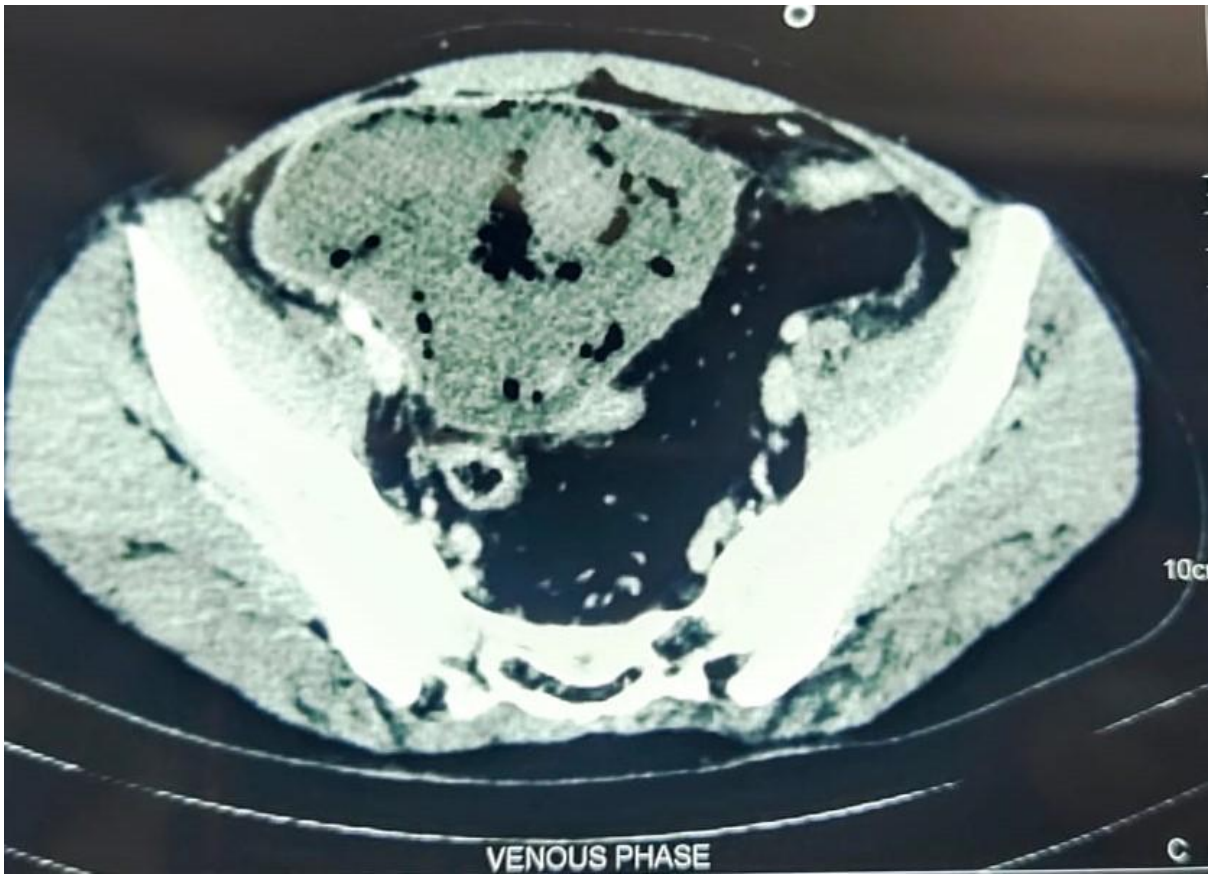

Fig 1 – CECT scan shows pocket of collection with internal air density

Regarding lab studies, there was leucocytosis with TLC 12000 [N 85 %], raised ESR, low serum albumin [2.5 gm/dl]. Serum urea was raised [93 mg/dl], serum creatinine was normal. Sodium slightly reduced with normal potassium level. These changes in urea and electrolytes were due to dehydration and sepsis.

Patient resuscitated with crystalloids, antibiotics started and decision taken in favour of emergency laparotomy.

On opening the abdomen, turbid, pus mixed fluid came out of which a sample was taken for culture & sensitivity. Thorough exploration done after suctioning and a gangrenous tumour like growth seen arising from the antimesenteric border of mid-jejunum. There was perforation on the tip of the mass. No other growth was seen and rest of the bowel was normal. Thorough peritoneal toileting done. In view of the proximal location of the lesion, making a stoma was ruled out. Wedge resection of the lump including healthy margin of jejunal wall was done and repaired in two layers. The specimen sent for histopathology. A segment of greater omentum also excised and sent for biopsy.

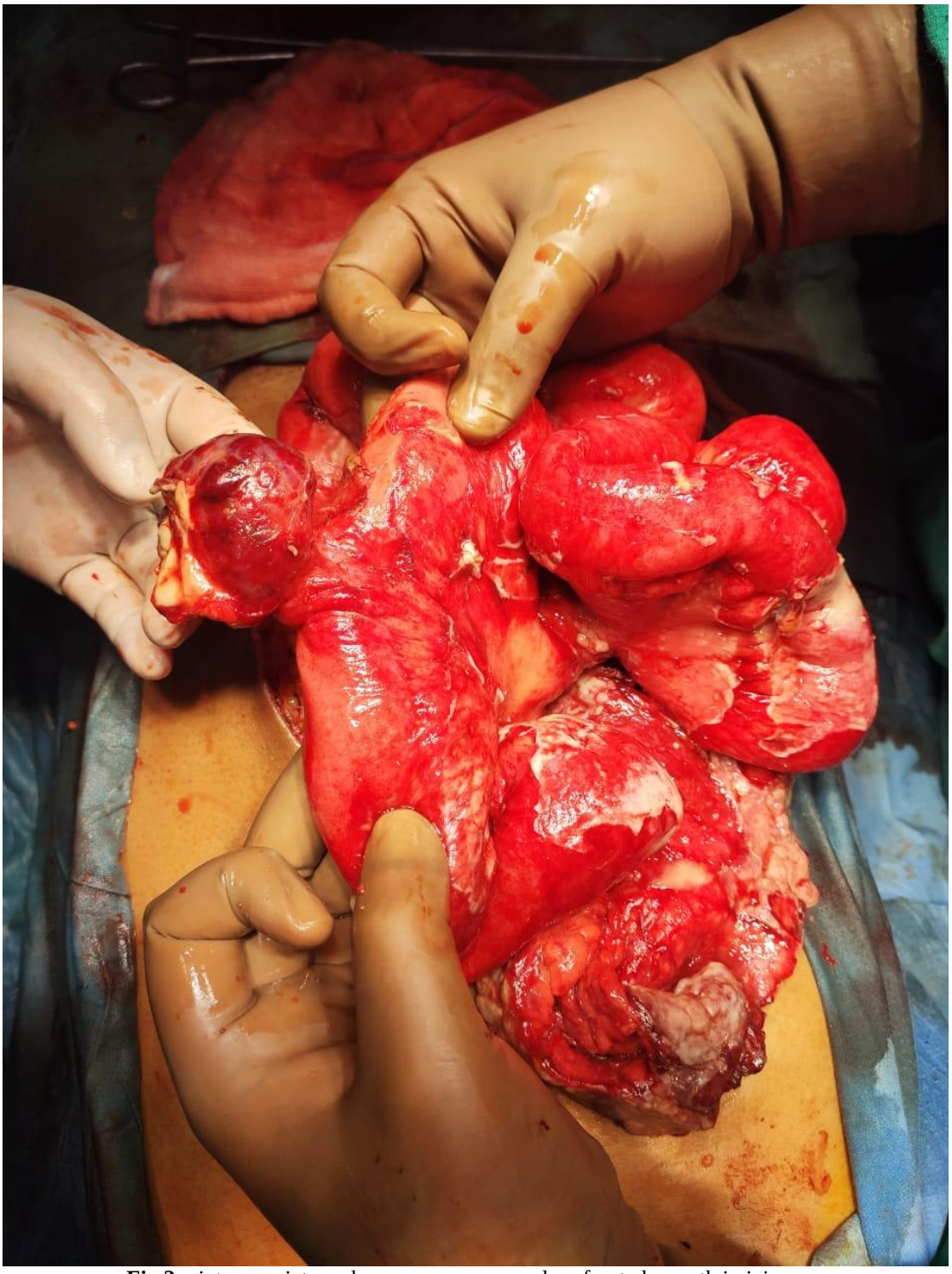

Fig 2 – intra op picture shows gangrenous and perforated growth in jejunum

Histopathological features were suggestive of poorly differentiated sarcoma with differential diagnosis of Gastro-intestinal stromal tumour or GIST, Rhabdomyosarcoma and amelanocytic myeloma.

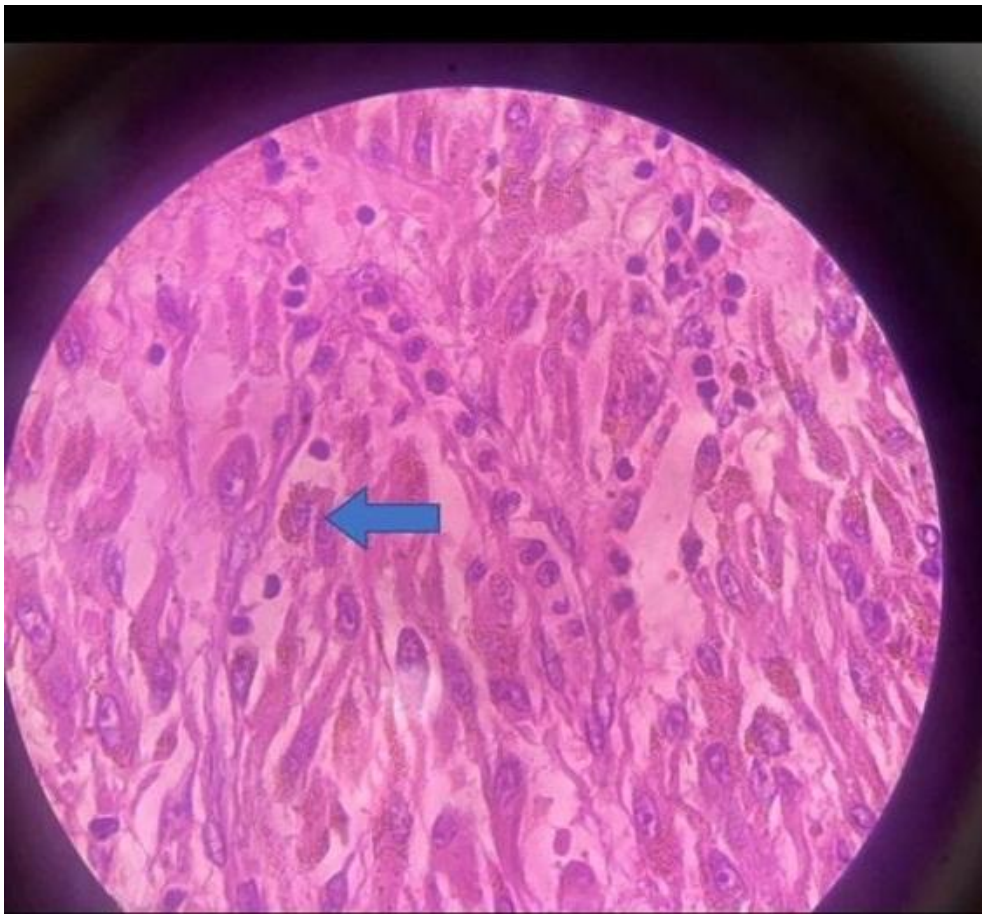

Fig 3 – tumour cells shows pigments

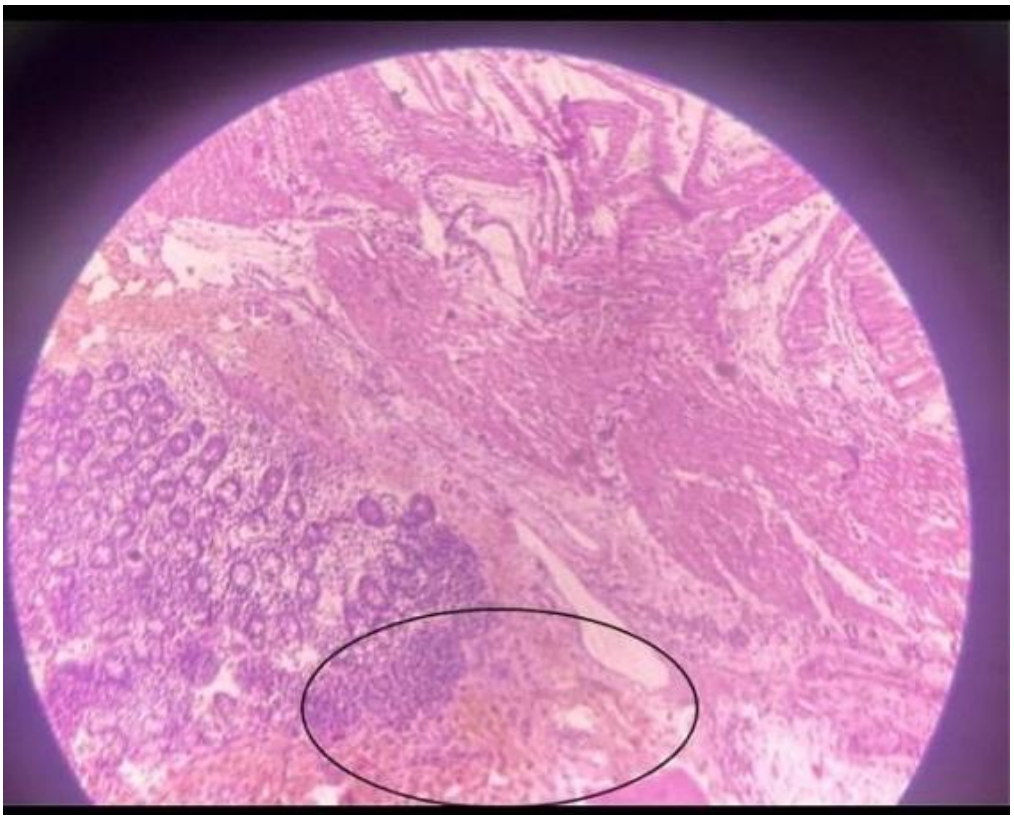

Fig 4 – section shows haemorrhage

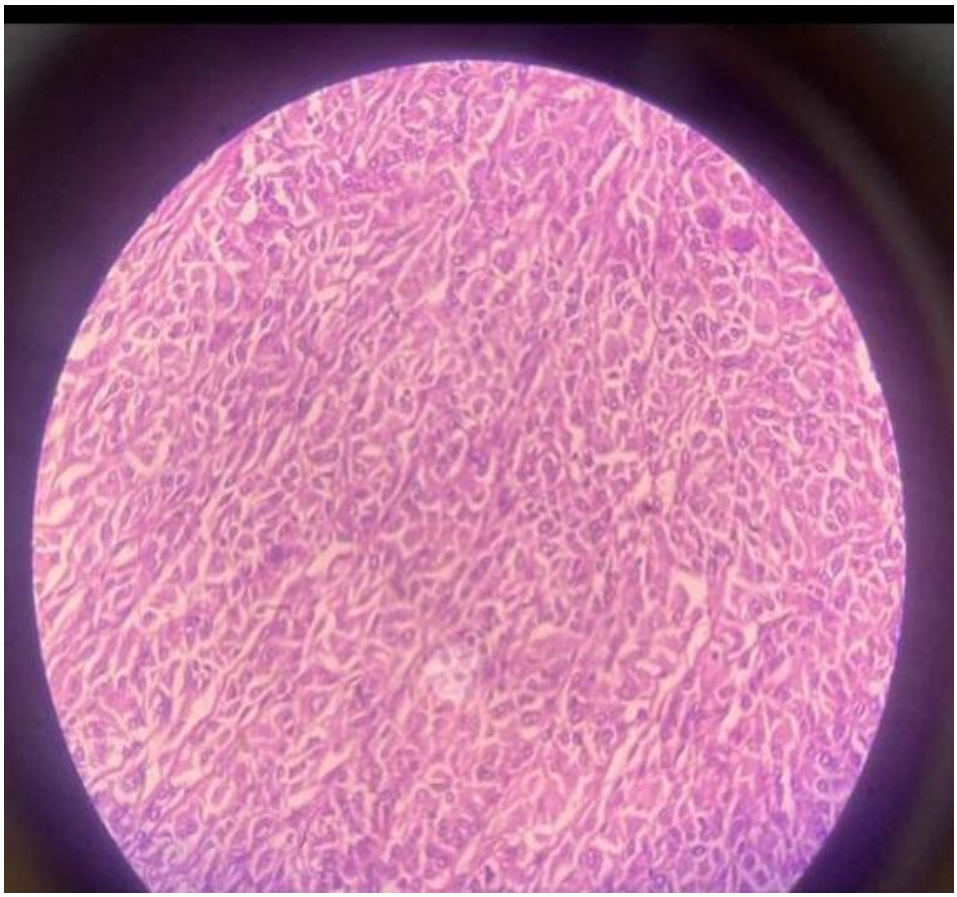

Fig 5 – section shows individual tumour cells

Figures 3, 4 & 5 shows various histological sections of the tumour. In view of the inconclusive histopathological report, it was decided to proceed with immunohistochemistry for confirmation of diagnosis.

Immunohistochemistry analysis shows CD 34 +ve, CD 117 +ve , Desmin –ve, HMB 45 –ve, Ki-67 -2-3% , S100 –ve, Smooth muscle actin –ve, Vimentin +ve, DOG1 +ve. Immunomorphology confirms diagnosis of "Epithelliod Gastrointestinal stromal tumour".

Post-operative recovery was uneventful except mild wound infection. Patient was prescribed Imatinib tablet 400 mg daily to continue for 3 years. Patient came for follow up after 3 months without any recurrence.

III. DISCUSSION

GIST was first described by Mazor and Clark (1983) [3].It originates from the interstitial cells of Cajal (ICC), located in the muscularispropria (myenteric plexus) responsible for triggering smooth muscle contraction [4, 5]. Thebasic pathology is an activating mutation (gain function) of chromosome 4 which codes for c-Kit resulting in uncontrolled proliferation of stem cells that differentiate towards ICC. GIST is sporadic [5]. Familial forms with autosomaldominant inheritance have also been documented [5,6]. Isolated reports of GIST occurring concomitantly with paraganglioma, pulmonary chondroma, nerofibromatosis, pancreatic neuro-endocrine tumours, burkitt's lymphoma, osteosarcoma, neuroblastomaand melanoma have been documented [6].

90% of GIST occurs in adults more than 40 years of age (median age 63 years). There is slight male preponderance [6]. No documented elements indicating any association with geographic location, ethnicity, race or occupation has been elucidated [6, 7].

GIST is a visceral tumour arising from any site of the gastrointestinal tract. Approximately 60-70% of cases occur in the stomach, 25-35% in the small intestine and 10% in the jejunum, while the oesophagus, colon, rectum and appendix are rarely affected [1]. Approximately 10-30% of patients with GISTs may be asymptomatic. The most common symptoms associated with jejunal GISTs are vague, non-specific abdominal pain or discomfort. Patients who have jejunal GIST usually suffer from abdominal pain or palpable mass, and also complain of early satiety or abdominal fullness. Jejunal GISTs may cause symptoms secondary to obstruction or haemorrhage. Pressure necrosis and ulceration of the overlying mucosa may cause gastrointestinal bleeding, and patients who experience significant blood loss may suffer from malaise and fatigue. Obstruction may result from the intraluminal growth of the tumour or luminal compression from an

exophytic lesion. Fever, anorexia and weight loss are rarely observed [2] and GISTs originating from the jejunum seldom cause perforation and acute diffuse peritonitis [3, 8-10].

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. Although a CT scan is a commonly offered imaging modality for patients with suspected abdominal GISTs, magnetic resonance imaging (MRI) provides better information than CT in the preoperative workup [11]. The definitive diagnosis of the majority of jejunal GISTs is revealed by histopathological examination of the specimen. Approximately 95% of GISTs express CD117, which is part of the KIT receptor tyrosine kinase. Additionally, DOG1, a recently defined monoclonal antibody against a chloride channel protein expressed by GIST, is positively expressed in 95% of GISTs [12]. DOG1 is a novel marker of GISTs as it has a higher sensitivity and specificity compared with CD34, particularly in the detection of moderate and high risk GIST.

In our case CT scan showed only signs of intraperitoneal collection and pneumoperitoneum suggestive of hollow viscous perforation. It failed to detectjejunal GIST pre-operatively. Even histopathology was not conclusive. Then immunohistochemistry showed both CD117 and DOG1 positivity, finally confirming the case as GIST.

To date, surgery is the only potentially curative therapy for patients with primary, resectable GIST. Nonmetastatic GISTs greater than 2 cm should be resected. A lymphadenectomy is not conducted because lymph node metastases are rare [13]. There were no positive lymph nodes in the present case. Partial omentectomy was done but no tumour cells were found in histopathological examination.

The management of GIST has undergone significant revolution over last decade. Tyrosine kinase inhibitor therapy has significantly improved overall survival in patients with advanced disease and should be continued indefinitely. Prior to the development of imatinib, recurrences were common even inpatients undergoing surgery. Adjuvant imatinib for 3 years should be considered in patients undergoing resection for primary disease [5].

IV. CONCLUSION

GIST is rare and its presentation as spontaneous rupture is extremely rare. Despite all the advancement in diagnostic procedures the preoperative diagnosis remains difficult. We report a case of a male with a perforated GIST in the jejunum causing acute diffuse peritonitis. The clinical outcome is worse when this tumour presents with bowel perforation and peritonitis. A high degree of suspicion is necessary in view of the high morbidity rates resulting from a delayed diagnosis of the disease.

Declarations :
Conflict of interest: None
Ethical approval: Not required

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