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Case Report

Spontaneous perforation of jejunal gastrointestinal stromal tumour presenting with multiple intraperitoneal abscess cavities: a case report and review of literature

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ABSTRACT

Gastrointestinal stromal tumours (GISTs) account for less than 1% of gastrointestinal tumours. Perforation of GIST is very rare (0.8%) and it is infrequently described in the literature. We report a case of 53 years old gentleman who had spontaneous perforation of jejunal GIST with multiple intraperitoneal abscess cavities which is unique and extremely rare.

Keywords: GIST, Perforation, Abscess

INTRODUCTION

Gastrointestinal stromal tumours (GISTs) account for less than 1% of gastrointestinal tumours, but they are the most common (80%) mesenchymal neoplasms of the gastrointestinal tract and can occur anywhere in the gastrointestinal tract.[1] Approximately 20 - 30% of GISTs originate from the jejunoileum. Up to 75% GISTs are either asymptomatic or associated with nonspecific symptoms. Perforation of GIST is very rare (0.8%) and it is infrequently described in the literature.[2,3] 15 – 47% of patients with GIST have metastasis disease at diagnosis.

CASE REPORT

We report a case of 53 years old gentleman who presented with black coloured stool since 5 days and pain abdomen associated with fever since 3 days. On admission, his vital signs were stable, while his physical examination revealed a palpable abdominal mass (Approx. 10×7 cm) in left lower quadrant. There was generalized tenderness (more in left lower quadrant). Bowel sounds were sluggish. He is a known case of type II diabetes mellitus (last 2 years) on regular medication. Plain X-ray (erect chest, PA view) showed no free gas under diaphragm. USG of whole abdomen suggested multiple intraperitoneal complex cystic space occupying lesions (average 6×5 cm) with free fluid in peritoneal cavity. CT scan of whole abdomen as given in Figure 1 suggested possibility of GIST. (Lobulated soft tissue lession 7×5×4 cm in the small bowel loops with central ulceration).

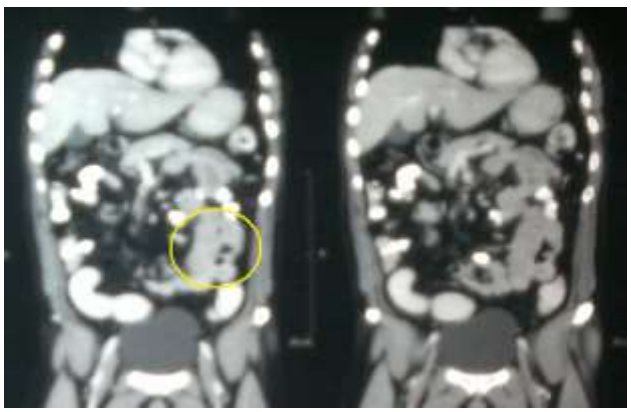

Figure 1: CECT of whole abdomen.

Haematological examination revealed moderate anaemia with neutrophilic leucocytosis (Hb 9.4 gm/dl, TLC 13,500/ cu.mm; N 85, L 11, E 10, B 0, M 0). Liver function test detected low albumin (2.2 gm/dl). Kidney function test was normal. Blood sugar (R)-124 gm/dl. After checking all baseline investigations exploratory laparotomy was done under general anaesthesia.

A 8×5×5 cm mass was noted in the distal jejunum. There were multiple perforations in the tumour mass as shown in Figure 2. Multiple intraperitoneal abscess cavities were present.

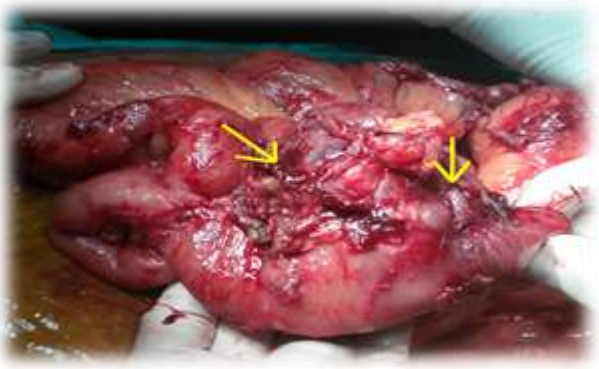

Figure 2: Intraoperative picture of perforated jejunal tumour mass (GIST).

No tumour mass or metastatic deposits were noted in other parts of bowel, peritoneum, liver or other organs. Peritoneal lavage, excision of tumour mass followed by end to end anastomosis of resected small bowel ends were done. Histopathology report confirmed it as gist of mixed spindle and epitheloid type with low mitotic activity (3/50 HPF) and areas of tumour necrosis as given in Figure 3. Tumor free margin was 7 cm and there was no lymph node involvement. Immunohistochemistry study revealed diffuse positivity for CD 117, focal positivity for CD 34 and negativity for desmin, S-100, SMA.

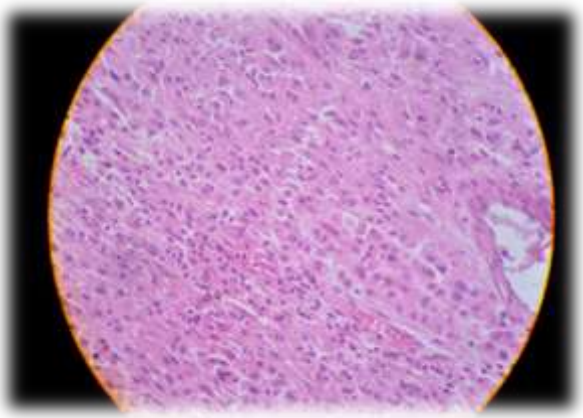

Figure 3: Histopathology slide of specimen (jejunal GIST).

TNM staging of our case was T3N0M0 (Stage II, 7th Edition *AJCC*). The patient was discharged on 10th postoperative day. He is currently receiving chemotherapy with Imatinib and no recurrence till now (1 year follow up).

DISCUSSION

GISTs predominantly occur in patients around the sixth decade of life and can be found in any site of gastrointestinal track with no significant difference in distribution between males and females.[7]

The symptoms and signs are not disease specific and as a consequence about 50% of GISTs have already metastases at the time of diagnosis, usually to the liver or the peritoneum.[8,9]

Although the diagnostic procedure includes several examinations, such as barium examination of the gastrointestinal track, CT and angiography, none of these can establish the correct diagnosis with 100% certainty.

The diagnosis of GIST may be suggested during surgery by the presence of a well defined extra-luminal mass. However, it always requires histological and immunohistochemical confirmation.

The treatment of GISTs is the surgical R0 resection of the primary tumor. Several reviews have reported that small GISTs (<2 cm) can be treated adequately by wedge (gastric) or segmental (bowel) resection. Larger GISTs may require more extensive resection including adjacent structures or organs if involved.[10]

There is no indication for chemotherapy and radiation therapy after surgical resection of GISTs as these tumors are notoriously unresponsive to such treatment. Radiotherapy is only used in cases of intraperitoneal hemorrhage, when the precise location of the tumor is known or for analgesic purposes.[11]

The molecular status of GISTs turns out to be relevant for the response to targeted treatment with Imatinib a powerful and relatively selective inhibitor of all ABL tyrosine kinases of platelet-derived growth factor receptor (PDGFR) and of c-kit receptor.

Optimal surgical treatment of GIST entails complete removal of the tumor with clear surgical margins including the adjacent involved organs.[12,13] Complete surgical resection entails 48-65% five-year survival.[2] Perforation of the tumor lowers the five-year survival to 24%, probably due to peritoneal dissemination.[12] Local and regional lymph node involvement is infrequent in GIST.[14,15] GIST's presenting with perforation, attention needs to be paid, in view of possible recurrence of the tumor. Abundant peritoneal lavage should be performed with distilled water to reduce the risk of peritoneal tumour spillage. Distilled water is used because of its

cytolytic activity on suspended cells.[16,17] The 5-year survival rate is 35%. It increases to 54% after complete surgical excision. However 40% will recur within 18 – 24 months. Once recurrence has occurred median survival is 9–16 months.[7,12,16]

Ulusan S et al. reported a case of ruptured GIST with pelvic abscess in a 52 years old woman.[18] To our knowledge, our case is the second published report of a spontaneously ruptured GIST of the small bowel presented with abscess cavities.

CONCLUSION

Although there are a few reports of perforated GISTs in the literature, spontaneous perforation of jejunal GIST with multiple intraperitoneal abscess cavities is unique and extremely rare.

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