

A Rare Case of Meckel's Diverticulum GIST presenting as Perforative Peritonitis

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Abstract : Tumours of Meckel's diverticulum are infrequent and account for 0.5 to 3.5% of cases. Of these, 12% tumours are GIST. A Meckel's diverticulum GIST presenting as Perforation is extremely rare¹. In English literature, we have found 8 well reported cases of Perforated Meckel's diverticulum GIST so far².

A 47 year old female patient presented as Acute Abdomen and was diagnosed as a case of Perforative Peritonitis. On Laparotomy, a Perforating tumour arising from Meckel's diverticulum was found, **Conclusion:** GIST is a KIT mutation driven mesenchymal neoplasm comprising about 0.1- 1% of all GI malignancies. Perforated GIST is associated with poor prognosis. Surgery is considered the standard treatment for non-metastatic GISTs with enbloc resection and clear margins³.

INTRODUCTION

Persistence of the intestinal end of the Omphalomesentric duct results in Meckel's diverticulum⁴. It is a true diverticulum in that it is composed of the same three layers that make up the ileum. In 20% of cases the innermost layer contains heterotopic mucosa⁵. The diverticulum is found in 2% of the population, 2 inches long and is situated on the anti-mesenteric border of the small intestine, commonly 2 feet from the ileocaecal valve⁶. It may be complicated by inflammation, perforation, haemorrhage, or obstruction. Meckel's diverticulitis is dangerous because its walls are thinner and it therefore perforates easily⁷. When perforation occurs, diffuse peritonitis follows quickly, and is more lethal than perforated appendicitis because the diverticulum is placed more centrally and there are fewer anatomical barriers to the rapid extravasations of liquid⁵. Much rarer complications of Meckel's diverticula include neoplasm's (0.5 – 3.2%)^{3,8-10}. Of these tumours 12% are GISTs^{3,8-9}. Meckel's diverticulum GIST presenting as perforation is extremely rare¹. So far one indexed case has been reported from India¹¹.

CASE REPORT

A 47 year old female admitted on 19/04/2014, presented with right lower abdominal pain of 3 days duration. On examination of abdomen, there was guarding and rigidity, more over right iliac fossa and hypogastrum. Diffuse tenderness was present all over the abdomen. No mass was palpable. Abdominal X-ray revealed pneumoperitoneum. A diagnosis of perforative peritonitis was made on 26/04/2014 and the patient was shifted for emergency laparotomy on the same day of diagnosis.

On laparotomy, there was about 600 ml of purulent fluid in right iliac fossa and hypogastrium wrapped with omentum along with perforated mass without contaminating the general peritoneal cavity. Purulent fluid sucked out and thorough lavage given after completing the surgery. A 7cm x 4cm proliferative growth was seen arising from a diverticulum in the ileum about 2 1/2 feet from ileocaecal junction in its anti-mesenteric border. About 1cm x 1cm perforation was seen in the growth, the perforation being continuous with bowel lumen. Wound got infected and allowed to heal by secondary intension. Patient discharged on 18/05/2014 and referred to medical oncology department for further management. Patient not turned up for further follow up.

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Histopathology revealed 4cm long small intestine with the intestinal wall showing a diverticulum and a 7cm x 4cm x 3cm mass arising from the diverticulum. Cut section showed greyish white mass with area of perforation. Microscopy revealed small intestine mucosa showing a diverticulum lined by ileal mucosa. The diverticular wall showed tumour tissue composed of spindle shaped tumour cells arranged in fascicles and sheets. There was no increase in Mitotic activity (< 2 per 50 HPF). The inference being GIST arising from Meckels diverticulum.

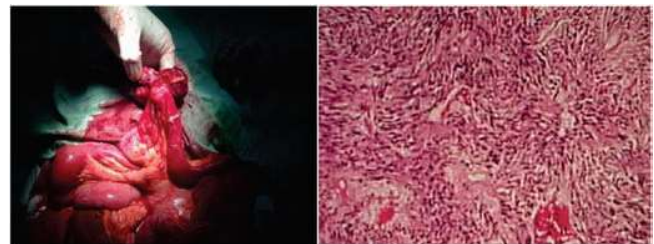


Fig. 1: Tumour in meckel's diverticulum **Fig. 2 :** Spindle shaped tumour cells arranged in fascicles and sheets

IHC –Report. KT Not done.

| | |
|----------|----------|
| C-kit | Positive |
| Vimentin | Positive |
| S100 | Negative |
| Desmin | Negative |
| Ki 67 | 3% |

Immunohistochemistry showed a C-kit positive and Desmin negative tumour confirming the diagnosis of GIST.

DISCUSSION

GIST is a KIT positive and KIT mutation driven mesenchymal neoplasm specific to the GI tract¹². GISTs are rare neoplasm's which account for 0.1–1% of gastrointestinal malignancies⁸. The majority of GISTs (60% to 70%) have been reported to arise in the stomach, whereas 20% to 30% originate in the small intestine, and less than 10% in the oesophagus, colon and rectum^{3,8}. GISTs also occur in the extra-intestinal abdominopelvic sites such as the omentum, mesentery, and retroperitoneum^{3,8}. GISTs arising from Meckel's diverticulum are extremely rare. They are now known to be derived from the interstitial cells of Cajal, an intestinal pacemaker cell¹². These cells have a stem- cell like character as demonstrated by their ability to transdifferentiate into smooth muscle¹².

Most GISTs approximately 85% - 90% contain oncogenic KIT or PDGFRA mutations¹². Approximately 90% of KIT mutations involve exon 11¹². Exon 9 mutations are rare and essentially restricted to intestinal GISTs¹². The most common presentation of GISTs is acute or chronic gastrointestinal bleeding³. Symptoms at presentation may include abdominal pain, abdominal mass, nausea, vomiting, anorexia, and weight loss. The preoperative diagnosis of GIST at this localization is difficult and is usually made during laparotomy and confirmed by histopathology on the excised material¹³. Pathologic diagnosis of GIST is based on identification of a mesenchymal neoplasm with spindle cell or epithelioid histology¹³. Common histologic features in GIST include spindle cells with sclerosing matrix, perinuclear vacuolisation and nuclear palisading¹³.

They are now frequently identified by immunohistochemical staining for the c-kit proto-oncogene (CD 117) found in more than 90% of these tumours and for CD 34, present in 80% of GISTs⁴. The most important adverse factors are thought to be a tumor diameter of greater than 5 cm and a high mitotic count exceeding 5 per 50 HPF on light microscopy³. The prognosis is dismal when the tumor presents with symptoms or signs such as perforation, multifocal location or metastatic lesions. Patients with localized or locally advanced tumors have 46% five-year survival, in contrast to patients with metastatic tumors or multifocal tumors in whom the five-year survival is 0%. Perforation of the tumor lowers the five-year survival to 24%, probably due to peritoneal dissemination¹⁴.

Segmental resection of tumor containing segment to obtain negative margins is the treatment of choice⁴. Wide resection of the mesentery with lymphadenectomy is not necessary as lymphatic metastasis is unusual⁴. Recurrence rates after resection were as high as 70%. Adjuvant Imatinib Mesylate is now the standard of care for malignant GISTs, especially with size larger than 5 cm, high mitotic rate or small bowel location⁴. Indefinite Imatinib is to be given for metastatic disease.

CONCLUSION

GIST of small bowel has traditionally been associated with poor prognosis. However, recent trials have shown that 1 year of adjuvant Imatinib mesylate after complete resection of a GIST, significantly improved recurrence free survival. Although preoperative diagnosis of GIST is difficult they should be considered in the differential diagnosis of small bowel tumours.

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LITERATURE REVIEW

ULTRASOUND AND MAGNETIC RESONANCE IMAGING CORRELATION OF THE WRIST AND METACARPOPHALANGEAL JOINTS IN FIFTY CONSECUTIVE PATIENTS OF RHEUMATOID ARTHRITIS

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Gray-scale ultrasound and Power Doppler ultrasound (GSUS, PDUS) and contrast Magnetic Resonance Imaging (MRI) are the imaging modalities to detect the disease in this early stage of rheumatoid arthritis (RA). This study was conducted to observe the advantages and disadvantages of USG and MRI of wrist and metacarpophalangeal joints in patients with rheumatoid arthritis. Clinically diagnosed cases of RA were included. GSUS-PDUS and high field (1.5T) MRI with contrast were used by two assessors. The evaluation and scoring was done using the RAMRIS score. Other assessed parameters included Joint space narrowing, GSUS bone erosions, Gray scale USG synovial hypertrophy, MRI tenosynovitis and tenosynovitis on GSUS Total 50 patients (44 female, 6 male; age 18-76 years) with 100 joints and 400 Metacarpophalangeal joints (2nd to 5th) including 2300 bone areas were evaluated. GSUS, PDUS evaluation was equal to contrast MRI evaluation in detecting joint space narrowing, effusion, flexor tenosynovitis (except for FPL tendon), extensor tenosynovitis. Whereas synovial thickening was better picked up in GSUS, PDUS than on contrast MRI. Contrast MRI picked up more cases of bone erosions, triangular fibro cartilage lesions and active synovitis as compared to GSUS, PDUS. Bone marrow edema which indicates active osteitis could only be directly detected on contrast MRI. **Conclusions:** GSUS, PDUS can be used as radiological investigative modality for the diagnosis of cases of early rheumatoid arthritis specially for synovial thickening, joint effusion, flexor and extensor tenosynovitis