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A RARE CASE OF MECKELS DIVERTICULUM GIST PRESENTING AS PERFORATIVE PERITONITIS

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Abstract : Background Tumours of Meckels diverticulum are infrequent and account for 0.5 to 3.5 of cases. Of these, 12 tumours are GIST. A Meckels diverticulum GIST presenting as Perforation is extremely rare11. In English literature, we have found 8 well reported cases of Perforated Meckels diverticulum GIST so far14. Case Presentation 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 Meckels 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 margins5.

Keyword :GIST, meckel's diverticulum, perforation

INTRODUCTION:

Persistence of the intestinal end of the Omphalomesentric duct results in Meckel's diverticulum1. 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 mucosa2. 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 valve3. It may be complicated by inflammation, perforation, hemorrhage, or obstruction. Meckel's diverticulitis is dangerous because its walls are thinner and it therefore perforates easily4. 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 extravasation of liquid2. Much rarer complications of Meckel's diverticula include neoplasms (0.5 – 3.2%)5,6,7,8. Of these tumours 12% are GISTs5,6,7. Meckel's diverticulum GIST presenting as perforation is extremely rare11. So far one indexed case has been reported from India9.

CASE REPORT:

A 47 year old female 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 hypogastrium. Diffuse tenderness was present all over the abdomen. No mass was palpable. Abdominal X-ray revealed pneumoperitoneum. A diagnosis of perforative peritonitis was made and the patient was shifted for emergency laparotomy. On laparotomy, there was about 600 ml of purulent fluid in peritoneal cavity. 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. Histopathology revealed 4cm long small intestine with the intestinal wall showing a diverticulum and a 7cmx4cmx3 cm 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 Meckel's diverticulum. Immunohistochemistry showed a C-kit positive and Desmin negative tumour confirming the diagnosis of GIST.

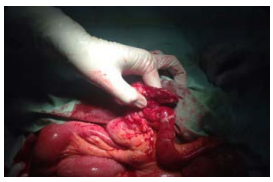
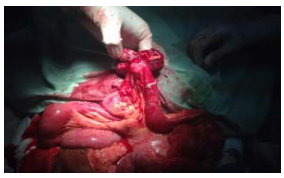


Fig1



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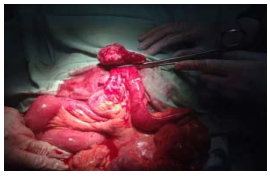


Fig3

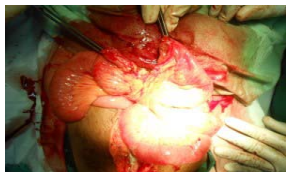
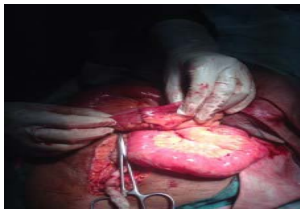


Fig4



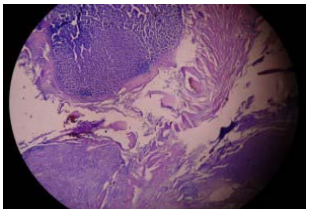
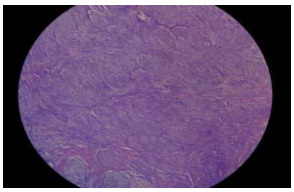


Fig5 **Fig6**



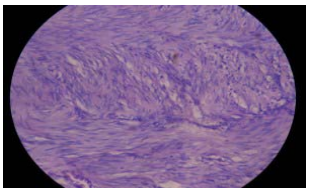


Fig7 **Fig8**

DISCUSSION:

GIST is a KIT positive and KIT mutation driven mesenchymal neoplasm specific to the GI tract 10. GISTs are rare neoplasms which account for 0.1–1% of gastrointestinal malignancies6. 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 rectum5,6. GISTs also occur in the extra-intestinal abdominopelvic sites such as the omentum, mesentery, and retroperitoneum5,6. 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 cell10. These cells have a stem- cell like character as demonstrated by their ability to transdifferentiate into smooth muscle10. Most GISTs approximately 85% - 90% contain oncogenic KIT or PDGFRA mutations10. Approximately 90% of KIT mutations involve exon 1110.Exon 9 mutations are rare and essentially restricted to intestinal GISTs10.The most common presentation of GISTs is acute or chronic gastrointestinal bleeding5. Symptoms at presentation may include abdominal pain, abdominal mass, nausea, vomiting, anorexia, and weight loss. The preoperative diagnosis of GIST at this localisation is difficult and is usually made during laparotomy and confirmed by histopathology on the excised material12. Pathologic diagnosis of GIST is based on identification of a mesenchymal neoplasm with spindle cell or epitheloid histology10. Common histologic features in GIST include spindle cells with sclerosing matrix, perinuclear vacuolisation and nuclear palisading10. They are now frequently identified by immunohistochemical staining for the c-kit protooncogene (CD 117) found in more than 90% of these tumours and for CD 34, present in 80% of GISTs1. 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 microscopy5. 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 dissemination13.

Segmental resection of tumour containing segment to obtain negative margins is the treatment of choice1. Wide resection of the mesentery with lymphadenectomy is not necessary as lymphatic metastasis are unusual1. 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 location1. 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 compete 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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