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CASE REPORT

ACUTE ABDOMEN PRESENTING AS MESENTERIC MALIGNANT GIST WITH PANCREATIC HETEROTOPIA IN THE JEJENUM – A RARE CASE REPORT

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ABSTRACT

Gastrointestinal stromal tumor (GIST) of alimentary tract has an incidence of about 1 per 100,000. Mesenteric GIST are rare and constitute less than 1 % of all gastrointestinal malignancies. Heterotopic pancreas in the jejunum is encountered incidentally and the incidence ranges around 0.2-0.9% of gastrointestinal surgeries. Here we present a rare case of 37 year old male who presented with acute intestinal obstruction and was operated immediately. On exploratory laparotomy, there were two masses, one in the mesentery and the other in the jejunum. Histologically the mesenteric mass was reported as malignant gastrointestinal stromal tumor and the jejunal mass was reported as heterotopic pancreas.

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INTRODUCTION

Gastrointestinal stromal tumors (GISTs) arise from the Interstitial cells of Cajal, and represent 1% of gastrointestinal tract malignancies. Primary mesenteric GIST are rare tumors constituting less than 1% of total cases of GIST. The diagnosis of GIST must be included in the differential diagnosis of any enlarging intrabdominal mass. Most GIST exhibit cytoplasmic immunoreactivity for cKIT (CD117). The patients are usually asymptomatic and it is often an incidental finding. It is a neoplasm occurring primarily in the gastrointestinal tract with age group >50 years. It rarely occurs outside the gastrointestinal tract in the vicinity of stomach or intestines, nearby areas such as omentum and mesentery. Heterotopic pancreas is pancreatic tissue developing in other areas and is usually asymptomatic malformation which can be present anywhere in the alimentary tract. Here we are presenting this case because the patient presented with symptoms of acute intestinal obstruction which was later confirmed histologically as gastrointestinal stromal tumor of the mesentery and the jejunal mass was reported as heterotopic pancreas an incidental finding on histopathology.

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CASE REPORT

37 years old male presented with sudden onset of abdominal pain since one day, history of vomiting and constipation since 15 days, and history of abdominal distension since two months. No significant past history/family history/surgical history. Patient was diagnosed as acute intestinal obstruction and laparotomy was done immediately.

Gross

Container labelled as mesenteric mass: Received a single gray white nodular mass measuring 5.5X5X4.5cm. E/S: Nodular. C/S: Grey white with whorled appearance.

Container B labelled as jejunal mass: Received two fragments measuring 3X1 and 2X1 cm. C/S: grey white.

Microscopy: Multiple sections from the mesenteric mass show highly cellular neoplasm composed predominantly of spindle cells arranged in interlacing fascicles and in storiform pattern. The cells have plump elongated nuclei, granular chromatin and scanty eosinophilic cytoplasm. In focal areas the tumor cells show palisading arrangement with wavy nuclei. Few foci shows epithelioid like cells arranged in sheets admixed with tumor giant cells. Atypical mitosis seen (>5/50 HPF) Immunohistochemistry showed CD117 positivity.



Figure 1. Gross specimen of A-mesenteric mass showing grey white firm areas and B- jejunal mass

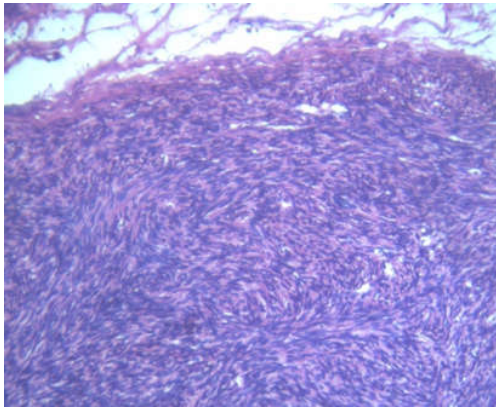


Figure 2. Low power view showing storiform pattern of spindle cells

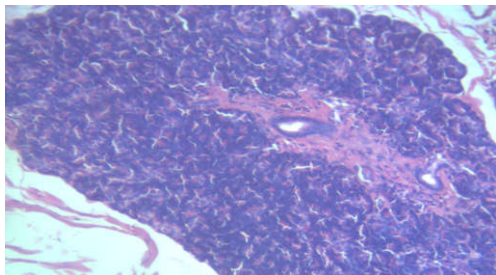


Figure 3. Low power view of jejunal mass showing pancreatic heterotopia

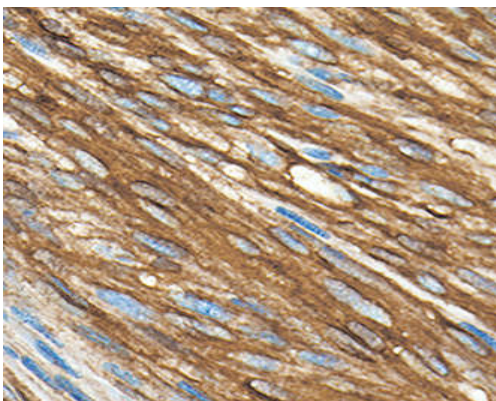


Figure 4. Immunohistochemistry positive for CD117

Sections studied from the jejunal fragments show lining by intestinal mucosa. The submucosa is occupied by heterotopic exocrine pancreatic tissue arranged in lobules of normally organized pancreatic acini and ducts. In focal areas the heterotopic pancreatic tissue is seen abutting on the mucosa. The histopathology showed, malignant gastrointestinal stromal tumor from the mesenteric mass confirmed by CD117 positivity and an incidental heterotopic exocrine pancreatic tissue.

DISCUSSION

GISTs are usually seen in the small bowel, followed by rectum and esophagus. They are rarely seen in the mesentery and constitutes less than 1 % of GISTs (Sinha *et al.*, 2004). The patient usually presents with pain abdomen (Feldman *et al.*, 2015). In our case patient presented with acute intestinal obstruction. Investigations done in our hospital showed large mass with incomplete resected margins, mitosis >5/50HPF and necrosis are all poor diagnostic factors. Surgery is the mainstay of treatment. Majority of GISTs are CD117 positive and few are PDGFRA positive (Puneet and Khanna, 2009). Heterotopic pancreas is a rare condition often diagnosed incidentally on histopathological examination and should be considered in the differential diagnosis of gastrointestinal lesions. Heterotopic pancreas usually lacks continuity with the main pancreas. It can occur anywhere in the gastrointestinal tract. Various theories of its origin has been put forward but it is usually an incidental finding intraoperatively. A definitive diagnosis of heterotopic pancreas is however made histopathologically. Heinrich's classification is commonly accepted, type 1: all elements of a normal pancreatic gland, type 2: pancreatic gland devoid of pancreatic islet cells, type 3: only pancreatic ducts are present. The preoperative diagnosis of heterotopic pancreas is difficult and the radiological findings does not distinguish other submucosal tumors with heterotopic pancreas. In our patient jejunal heterotopic pancreas was detected incidentally intraoperatively along with gastrointestinal stromal tumor.

Conclusion

Mesenteric GIST with jejunal heterotopic pancreas in the same patient is a rare incidental finding. Mesenteric GIST can be misdiagnosed as mesenteric cyst and can present as an acute abdomen. In any case with short duration of abdominal masses the differential of GIST has to be ruled out.

Conflict of interest: Nil

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