Abstract:
Introduction: Jejunal diverticulosis is an uncommon condition and its association with gastrointestinal stromal tumor (GIST) is very rare. We report a rare case of GIST arising in a jejunal diverticulum presenting as a pelvic mass. Case report: A fifty year old lady presented with a mass in the right lower abdomen associated with pain, vomiting and loss of weight. Computerised tomography of the abdomen showed a large, heterogenous mass arising from the right ovary. At laparotomy, however, she was found to have a large lobulated tumor arising from the jejunum which was resected. The mucosal surface of the resected jejunum showed the mouth of the diverticulum harbouring the tumor. Histopathology was suggestive of high grade GIST which was confirmed by immunohistochemistry. Patient was started on Tab. Imatinib 400 mg/day. Conclusion: Abdominal cavity continued to be a pandoras box and despite improvised imaging studies like computerized tomography, laparotomy is the final answer in clinching the diagnosis. Gastrointestinal stromal tumor (GIST) in a true jejunal diverticula is extremely rare and can be confirmed by immunohistochemistry. Keyword: Small bowel GIST, Jejunal diverticulum, Gastrointestinal stromal tumor

Case report:

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A fifty year old lady presented with right lower abdominal mass of 6 months duration associated with occasional dull aching pain and vomiting. She had significant loss of weight and appetite. She had no history of fever, gastrointestinal bleed or altered bowel habits. On clinical examination, there was a 18x10 cm non tender firm mass occupying the right iliac and hypogastric regions extending into the pelvis. Per rectal examination was normal. Rest of the systemic examination was unremarkable. Contrast enhanced Computerised Tomogram of the abdomen and pelvis showed a large lobulated heterogenous mass lesion arising from the right ovary; suggestive of a malignant neoplasm (Figure 1). Her blood investigations were within normal limits. At laparotomy, however, she was found to have a 20 x 15 cm lobulated encapsulated tumor arising from the antimesenteric border of jejunum, 30 cms from the duodenojejunal flexure (Figure 2), which was resected with a margin of 5 cms. There were 2 other jejunal diverticula proximal to the mass which were not resected. The fallopian tubes and ovaries were normal. On sectioning the jejunum, the mucosal surface revealed a diverticulum leading onto the mass (Figure 3). Histopathological examination was reported as spindle cell gastrointestinal stromal tumor arising from the antimesenteric border of jejunum, 30 cms from the duodenojejunal flexure (Figure 2), which was resected with a margin of 5 cms. There were 2 other jejunal diverticula proximal to the mass which were not resected. The fallopian tubes and ovaries were normal. On sectioning the jejunum, the mucosal surface revealed a diverticulum leading onto the mass (Figure 3). Histopathological examination was reported as spindle cell gastrointestinal stromal tumor (21cms) arising in a jejunal diverticulum (Figure 4). On immunohistochemistry, the tumor cells were positive for CD 117 (Figure 5). Mitotic activity was 3-4/50 high power field. It was graded as a high risk GIST in view of its size. She had an uneventful recovery and was started on adjuvant Tab. Imatinib 400mg /day.

Discussion:
Diverticulum can occur anywhere from the esophagus to the rectosigmoid. Small bowel diverticulosis is infrequent and is often detected incidentally on laparoscopy, laparotomy, radiographically or at post mortem. Incidence of jejunal diverticulosis on autopsy is 0.06-1.3% and 0-1.25% on radiography. Majority (80%) of these diverticula occur after the age of 40 years. Most of these diverticula are false diverticula arising from the mesenteric side of the bowel as a result of the mucosal herniation through the point of entry of blood vessels. True diverticulum comprising of all 4 layers of bowel are rare and present on the antimesenteric border. Only 50% percent of patients with jejunal diverticula are symptomatic. Symptoms occur secondary to bacterial overgrowth, inflammation or bleed within a diverticulum. They can also present with mechanical obstruction, perforation or can produce a malabsorption syndrome. Tumors within jejunal diverticula presenting as a lower abdominal mass are rare and have not been reported in literature. Sadaf et al reported a case of GIST in a jejunal diverticulum with obscure gastrointestinal bleed whereas Schepers et al reported a patient with cramping abdominal pain and diarrhea secondary to GIST in a jejunal diverticulum.

GIST is the most common mesenchymal tumor of the gastrointestinal tract, yet their incidence is 0.1-1% of all gastrointestinal malignancies. Small bowel is the second most common location for GIST comprising nearly 20-30%. They are believed to arise from the interstitial cells of Cajal, which are components of the intestinal autonomic nervous system that serve as pacemakers regulating intestinal peristalsis. These tumors are associated with a mutation in the tyrosine kinase c-kit oncogene or PDGFRA gene, and may or may not stain positively for kit. Ninety five
percent of all GISTs are CD 117 positive. The other markers include CD 34, DOG -18, desmin and vimentin.

Surgery is the definitive therapy for patients with GIST. Radical and complete surgical excision offers the only chance for cure. The most reliable prognostic factors are the size of the primary tumor and mitotic index which measure proliferative activity of the cells. The site of the primary tumor also has a bearing on the recurrence and survival rates, with small bowel tumors showing a somewhat worse prognosis. All high grade and inoperable tumors require treatment with Imatinib to downstage them. Tumors with mutations in exon 11 of c-kit are particularly sensitive. Our patient did not have a pre operative diagnosis of GIST and hence was taken up for laparotomy and was found to have a high grade GIST arising from the jejunal diverticula. As per the NCCN guidelines, she was started on Tab. Imatinib 400 mg per day.

Conclusion:
Abdominal cavity continues to be a pandora’s box and despite improvised imaging studies like computerized tomography, laparotomy is the final answer in clinching the diagnosis in some cases. Gastrointestinal stromal tumor (GIST) in a true jejunal diverticula is extremely rare. It can present as an intra abdominal mass and can descend into pelvis due to gravity delaying presentation to general surgeons.

Bibliography:


9. Christopher Kosmidis, Christopher Efthimiadis, Sofia Levva, George Anthimidis, Sofia Baka, Marios Grigoriou etal. Synchronous colorectal adenocarcinoma and gastrointestinal stromal tumor in Meckel's
Figure 2: Diverticulum harbouring the GIST

Figure 3: Jejunal mucosa showing the mouth of the diverticulum

Figure 4: H & E stain shows a spindle cell GIST

Figure 5: CD 117 positivity on immunohistochemistry