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General Surgery

A RARE CASE OF JEJUNAL GROWTH PERFORATION – HISTOPATHOLOGICAL SURPRISE

Dr. K. Arun Guru Assistant Professor Department of general surgery Velammal medical college and research institute Madurai

Dr. M. Karthika* Post Graduate Department of general surgery Velammal medical college hospital and research institute *Corresponding Author

ABSTRACT Gastrointestinal stromal tumors (GIST) are mesenchymal malignancies of the digestive tract, which mainly affect the stomach and small intestine. Jejunum is one of uncommon site for GIST and perforated jejunal GIST is one of rare complication .In the present study we report a case of a 38-year-old female presented with perforative peritonitis and laparotomy revealed jejunal small bowel perforation and biopsy confirmed as GIST. In this paper, we report the clinical manifestation as well as computed tomography and histopathological findings helpful for the accurate diagnosis of this rare complication of GIST. Emergency laparotomy and complete resection of tumor are essential. Specimen should be sent for immunohistochemistry for confirmation and tyrosine kinase inhibitors bsuch as imatinib should be given as adjuvant treatment

KEYWORDS : gastrointestinal stromal tumor (GIST), perforation, small intestine, Jejunum

Introduction

Gastrointestinal stromal tumors (GIST) are mesenchymal tumors of the digestive tract that originate from interstitial cells of cajal and account for 0.1–3% of all gastrointestinal tumors. They are usually located in the stomach and small intestine but they can be located anywhere in the gastrointestinal tract, including the omentum and peritoneum. Generally, GISTs have a silent behavior and are diagnosed incidentally. Approximately 40% of GIST cases cause intestinal bleeding Perforation is rarely observed in GISTs; however, we present a case of perforated GIST located in the jejunum as a rare cause of acute abdomen.

CASE REPORT

- 38 years old female presented to Emergency department with of abdomen pain, vo symptoms miting, low grade fever and abdominal distension for past 1 day. Per abdominal findings revealed distended abdomen with tenderness over epigastrium, right and left hypochondrial regions. Guarding and rigidity noted over upper abdomen. Bowel sounds were sluggish.
- Contrast enhanced CT Abdomen images showed Diffuse submucosal edema with wall thickening in mid jejunum and mucosal hyperenhancement with features of Perforation peritonitis. The decision was made to undertake an Emergency exploratory Laparotomy in view of perforated hollow viscus. After adequate resuscitation and preop workup, Patient was shifted to OT.
- Laparotomy was performed which revealed 4*3 cm exophytic growth over antimesentric border of jejunum approximately 5 cms from DJ flexure which was perforated with biliary peritonitis. The perforated part along with the exophytic growth was resected with Primary closure of jejunum in 2 layers.
- Surprisingly, histopathological examination revealed the diagnosis of GIST. Mitotic activity was 4-5 per 50 hpf . On immunohistochemistry, the tumor was strongly positive for CD117 and DOG-1 while negative for CD34, S-100 and SMA. Patient was started on oral imatinib 400mg once daily and was on follow up for the past 5 months.

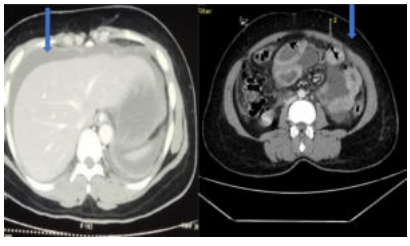


Figure 1. CT images showing perforated peritonitis with jejunal wall enhancement and free fluid in abdomen

Figure2. Intraop images showing jejunal growth perforation and resection with clear margins

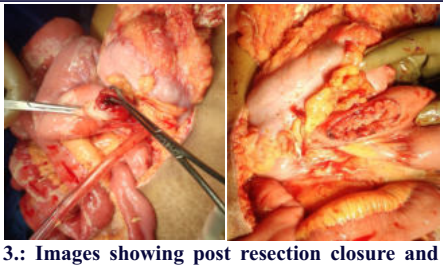


Figure 3.: Images showing post resection closure and resected specimen

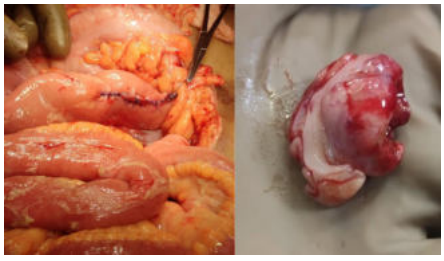
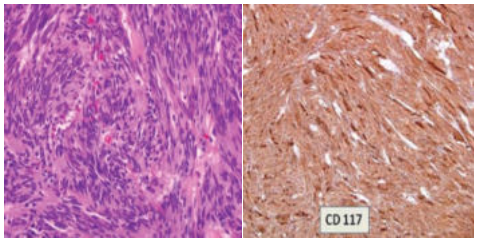


Figure 4. Histopathological slide and immunohistochemistry positive for GIST



DISCUSSION :

- GISTs account for <1% of Gastrointestinal tumours. GISTs arise from interstitial cells of CAJAL. They are the pacemaker cells of GI tract which are responsible for initiation of peristalsis. The stomach (65%) is the commonest site for GISTs followed by small intestine (30%), colon/rectum (5%) and esophagus (<1%). Generally, GISTs are benign in 70–80% of cases, particularly those located in the stomach.

- The most common clinical presentation of GIST is abdominal pain or GI bleeding, which manifests either chronically as anemia or acutely as melena or hematemesis. Other presentations include intestinal obstruction, nausea, weight loss, palpable mass and rarely perforation. Delayed presentation can also occur. GISTs under 2cm are often asymptomatic and when symptoms occur, they tend to be non-specific. 50% of malignant GISTs are therefore metastatic at the time of diagnosis.

- Histologically, GISTs consist of spindle, epithelioid or pleomorphic tissue. It can be confirmed immunohistochemically via staining for KIT protein (CD117). However, some tumours have PDGFRA mutations instead, rendering them CD117 negative. In this instance, DOG1 is a useful surrogate marker.

- Surgical resection is the mainstay of treatment for GISTs. Further treatment options are based on prognostic factors which include age at presentation, anatomic location, tumour size, mitotic rate, histomorphology, immunohistochemistry and molecular genetics.

- Adjuvant chemotherapy in the form of imatinib, a tyrosine kinase inhibitor results in prolonged survival rates in cases of advanced or metastatic disease. In cases of disease progression on imatinib, a second-line tyrosine kinase inhibitors such as sunitinib, ponatinib and regorafenib can also be used.

CONCLUSION :

- To conclude, GIST is a rare tumor that accounts for a small percentage of gastrointestinal neoplasms. Small intestinal GISTs and its presentation as Spontaneous perforation is extremely rare. Despite all the advancement in diagnostic procedures, preoperative diagnosis still remains difficult. However, Surgical resection of GIST with adjuvant imatinib therapy is the gold standard for the treatment of GISTs. If the tumor is unresectable, then neoadjuvant therapy with imatinib followed by resection is recommended.

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