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Case Report

A rare case of giant GIST: a case report and review of literature

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ABSTRACT

Gastrointestinal stromal tumors are very rare tumors that constitute less than 1 percent of entire tumors of GIT. The case reported here is a 60 years male who came with complaints of mass in the abdomen and constipation. On examination, he had guarding. Radiological evaluation revealed a heterogenous mass behind the lesser sac displacing the solid organs posteriorly. And hence we proceeded with total gastrectomy with esophago-jejunostomy. Histopathological examination revealed GIST arising from stomach which was positive for CD34 and C-KIT. Post-surgery patient was started on adjuvant imatinib therapy.

Keywords: Gastrointestinal stromal tumour, Imatinib, Sunitinib, CD34, CD117, DOG-1, Gastric GIST, PDGFR-A

INTRODUCTION

Gastrointestinal stromal tumours (GISTs) are mesenchymal tumours originating primarily in the gastrointestinal tract (GIT), with the stomach being the most commonly affected site (56%), followed by the small intestine (32%), colon and rectum (6%), and oesophagus (<1%).[1]

Typically, GISTs are smaller, usually not exceeding 5 cm in their largest dimension, although cases of Giant GISTs have been documented in the literature. These tumours are rare, with an incidence of 10-15 cases per million populations.[1] Here, we presented a case of a significant management challenge posed by a large GIST at our institution.

The largest GIST reported in the literature measured 42 cm×31 cm×23 cm and originated from the stomach.[2] The mass resected at our institution measured 32.5 cm×26.5 cm×15.5 cm, underscoring the substantial size and complexity of this particular tumour.

CASE REPORT

A 60-year-old male presented with a week-long history of upper abdominal pain, vomiting, and dull, continuous discomfort, accompanied by early satiety, heartburn, constipation, weight loss, and diminished appetite. There were no reports of melena, hematemesis, or jaundice. Physical examination revealed a distended abdomen with an ill-defined mass in the epigastrium extending into the right and left hypochondrium, exhibiting tenderness and guarding.

Further investigation through contrast-enhanced computed tomography (CECT) of the abdomen identified a sizable mass measuring 20×22×10 cm in the upper abdomen, exerting pressure on the stomach. Due to obstructive symptoms, surgical intervention was deemed necessary. Intra-operatively, the mass was found to originate from the body and fundus of the stomach, completely occupying the upper abdominal cavity.

Given the inability to establish a proximal margin on the stomach, a decision was made to perform a total

gastrectomy with oesophago jejunostomy, in addition to a feeding jejunostomy. The postoperative course was uneventful, and the patient was discharged on postoperative day 10.

Subsequent histopathological examination revealed a mass measuring 32.5×26.5×15.5 cm with 50% necrosis originating from the stomach. It was characterized as a low-grade spindle cell type gastrointestinal stromal tumour (GIST), positive for CD-34 and C-KIT.

DISCUSSION

GISTs once categorized as smooth muscle tumours of the gastrointestinal tract (GIT), underwent reclassification in 1983 upon the discovery that they originate from the interstitial cells of Cajal, pivotal in intestinal peristalsis and located within the circular muscles of the GIT. Predominantly positive for KIT Tyrosine kinase receptors (CD117) and PDGFR-A mutation, GISTs also express CD34 (70%), SMA (30 to 40%), Desmin (<5%), S100 (~5%), BRAF, and DOG-1.[3]

While GISTs are often asymptomatic, larger tumours may manifest with abdominal lump and other abdominal symptoms. Approximately 21% are incidentally discovered during surgery, with 10% found during autopsy.[4] In some cases, GISTs can lead to life-threatening complications such as hemorrhage into the bowel or tumour rupture causing hemoperitoneum.[5]

The main prognostic factors for GISTs include tumour size, mitotic index, and site of origin. Mitotic count, in particular, holds paramount importance in determining the malignant potential of the tumour.[6] Studies have indicated that small intestinal GISTs and those with more than 5 mitoses per 50 high-power fields (HPF) are associated with reduced overall survival. Additionally, poor prognosis from a genetic standpoint is linked to mutations such as aneuploidy, exon 9, and exon 11 deletion.[7,8]

Surgery stands as the cornerstone of GIST treatment, typically aiming for a resection margin of 1 to 2 cm, as preferred.[1] Following NCCN guidelines, tumour resection with histologically negative margins is advocated, usually achievable through segmental or wedge resections to secure negative margins, with extensive anatomical resections seldom necessary. Due to the delicate nature of GISTs, precautions must be taken to prevent tumour rupture, spillage, or laceration during surgical manipulation.

Minimally invasive surgeries are feasible for masses up to 5 cm.[10] Decisions regarding upfront surgery are guided by the extent of morbidity associated with resection. In cases of larger or resectable GISTs accompanied by significant morbidity, molecular testing and Immunohistochemistry studies are conducted, allowing for planned neoadjuvant targeted therapies before resection.[9] Adjuvant therapy, primarily through the tyrosine kinase inhibitor imatinib, is administered for a duration of 3 years, resulting in improved overall survival and recurrence-free survival compared to a 1-year regimen.[5]

Cases resistant to Imatinib, particularly those with Exon-18 mutations, may necessitate treatment with avapritinib.[11] Sunitinib is commonly employed as a second-line drug, followed by Regorafenib as a third-line option. In instances where GIST proves resistant to all three drugs, Ripretinib may be prescribed. Additionally, mTOR inhibitors like Everolimus and Kit transcription inhibitors such as Fluvopiridol are among the alternative treatment modalities utilized.

CONCLUSION

The primary treatment goal is achieving surgical resection with negative margins. For larger GISTs, preoperative systemic therapy may be considered to downsize the tumour and enhance operability. In this case surgery was considered as primary treatment as patient had obstructive features. On following up the patient, imatinib oral therapy was started and is taking the drugs.

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