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Dual Tumors Of GIT: Synchronous Neuroendocrine tumour of the Small Intestine and Gastrointestinal stromal tumour of the stomach - An interesting case report

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Abstract

Introduction

Gastrointestinal stromal tumours (GISTs), which stem from interstitial cells of Cajal located within the wall of the gastrointestinal tract and have a characteristic immunoreactivity for CD117 (c-kit protein), account for the majority of gastrointestinal mesenchymal neoplasms. It comprises of less than 2% of all gastrointestinal tumors. Its association with tumor of different histology is quite rare. Neuroendocrine tumors (NET) of the gastro-enter pancreatic system are also rare, representing about 2% of all gastrointestinal neoplasms. Synchronous occurrence of a GIST with a NET in the small intestine is extremely rare. In this case report author wants to report occurrence of synchronous tumor of the GIT (GIST of the stomach and NET of Small intestine).

Case Report

An elderly lady who presented to the ED of R. L. Jalappa Hospital with complaint of pain abdomen, vomiting and constipation since 3 days. Patient was dehydrated. Patient had distended abdomen with diffuse tenderness, guarding and rigidity. DRE was not contributory. Provisionally patient was diagnosed as acute intestinal obstruction.

Patient had undergone routine blood investigations which was within normal. Erect abdomen was taken which showed dilated bowel loops with multiple air fluid levels in step ladder fashion.

Patient had undergone emergency exploratory laparo to my. Intra operatively dilated proximal bowel with ileal stricture present 100 cm distal to DJ flexure. Stricture was circumferential and 2cms in length. Resection and anastomosis of Ileal stricture was done. Incidentally a Sessile exophytic growth was noted in the distal portion of stomach along greater curvature of 5x5 cm size.

Wedge resection of growth was done. Specimen was sent for histopathological evaluation. Post operative period was uneventful.

Keywords: Gastro intestinal stromal tumors, Neuro endocrine tumors, synchronous tumor.

Figure 1: Per op picture of Stricture in proximal ileum

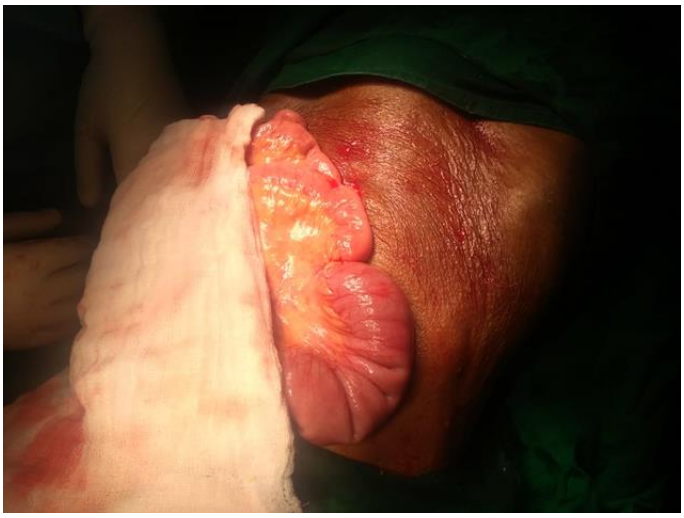
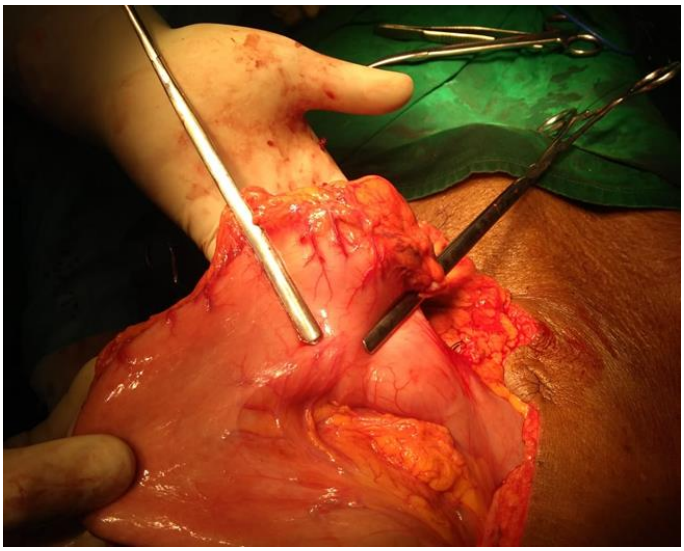


Figure 2: Per op picture of Exophytic growth arising from stomach



Histopathology

Histopathological report from Ileal specimen showed grey white lesion at stricture site with salt and pepper chromatin and scanty cytoplasm extending up to serosa. Proximal and distal segment are unremarkable. Features suggestive of Neuroendocrine tumor in intestine- Grade I

Histopathological report from resected stomach specimen showed

spindle shaped cells having elongated uniform nuclei with indistinct cytoplasm arranged in fasciles with focal myxoid changes Mitosis <2/10HPF- features suggestive of GIST- tumor of uncertain malignant potential.

Discussion

GIST one of the common mesenchymal tumors of GI tract encountered in routine practice. The mode of presentation varied from asymptomatic to ruptured GIST mimicking a acute abdomen. The order of frequency of GIST are like stomach (50-62%) > small intestine (20-30%) > colon (11%) > the rectum (7%). These tumors are associated with gain-of-function mutations in exon 11 of c-kit protooncogene.

Neuroendocrine tumors which arise from neural crest cell derivative i.e enterochromaffin cells (Kulchitsky cells) usually occurs in the 7th decade of life. The most common site for the NET is Lungs followed by bronchi and GIT(Appendix> small intestine (proximal ileum). These tumors are Categorized into Low grade, Intermediate grade and High-grade tumor based on their mitotic rates, invasion and Ki-67 proliferative index.

These tumors generally occur and manifest as a single tumor but there are few cases which was reported where they may either as synchronous dual or triple tumor. The cause for the occurrence of the synchronous tumors are still unsolved. Few studies had postulated the common carcinogenic pathways or genetic mutations with proliferation of different cell lines as the cause of such synchronous dual tumor.

Most of these tumors are asymptomatic or diagnosed incidentally. It may mimick like a acute abdomen and present like features of the intestinal obstruction or peritonitis. So in such circumstances a careful analysis

during surgery and its correlation with histopathological report helps us in arriving at accurate diagnosis.

Surgery is the most definitive therapy for patients with GIST which offers a better chance for cure. Surgery is even indicated for patients with locally advanced or metastatic disease e.g. debulking of the large lesions is helpful when adjuvant therapy is contemplated. Other option available in the line of management is chemotherapy. The drug which is quite often used is Imatinib which is a tyrosine kinase inhibitor which is recommended for minimum of 1 year after surgery and 3 years in case of recurrent tumors.

Similar to the GIST, surgery is the first-line therapy for NET too. And surgical option varies from resection and anastomosis to tumor bulking in case of locally advanced and metastatic tumor as it helps in decreasing the tumor mass thus low dose of chemotherapy will be required subsequently.

In the case reported, author had done resection anastomosis for the NET of small intestine and wedge resection of the stomach followed by adjuvant chemotherapy with imatinib.

Conclusion

The author wants to conclude that for acute abdomen cases possibility of the ruptured tumor should be kept as one of the differential diagnosis considering the patient profile. Resection of the tumor with oncological clearance should be the target even if done in acute setting as it gives a very good outcome as seen in our case, but the only condition for this is hemodynamically stable patient.

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