CASE REPORT

A 60 year old Hindu male presented in outpatient department of General Surgery with chief complaints of constipation since 4 month and pain abdomen in left lower quadrant since 2 month, relieved during surgery hence primary anastomosis of large and small bowel was not done. Postoperative period was uneventful and patient discharged on day 7 with stable vitals, with functional ileostomy and healthy suture line. Histopathological Report of multiple sections from

Extra Gastrointestinal Tumour (E-GIST): A Rare Case of Pelvic GIST
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Abstract: Gastrointestinal stromal tumors (GISTs) are rare tumors that can arise anywhere in the tubular gastrointestinal tract. A 60-year-old male presented with a pelvic mass measuring 12×10cm in diameter. He underwent exploratory laparotomy with excision of the pelvic mass. The mass was found in the pelvis adherent to bowel loop and sigmoid colon, was correctly diagnosed as a Extra Gastrointestinal stromal tumor (E- GIST) at surgery. It should be noted that preoperative diagnosis of E-GIST is uncommon, due to its rarity and the varying clinical presentation. Gynecologists need to be cognizant of extra-ovarian pathology in female patients presenting with an atypical pelvic mass.

Keywords: Extra gastrointestinal stromal tumors (E-GISTs), pelvic mass, sigmoid colon.
pelvic mass revealed Spindle cell Mesenchymal neoplasm, Mild to Moderate Anaplasia with Mitotic activity and area of necrosis. Sections from adherent segment of ileum and colon showed congestion and chronic inflammation with fragments of mass adherent on serosal surface. On follow up, tumour marker study showed following findings (Table 1).

Table 1: Immunohistochemistry (IHC) report

<table>
<thead>
<tr>
<th>IHC Marker</th>
<th>Result</th>
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</thead>
<tbody>
<tr>
<td>CD117/c-KIT</td>
<td>Positive</td>
</tr>
<tr>
<td>CD 34</td>
<td>Positive</td>
</tr>
<tr>
<td>Smooth Muscle Actin</td>
<td>Positive</td>
</tr>
<tr>
<td>KI-67</td>
<td>Positive (3-4%)</td>
</tr>
<tr>
<td>S – 100 P</td>
<td>Positive</td>
</tr>
</tbody>
</table>

After 2 months, patient was taken for Laprotomy for maintaining large bowel continuity by using circular stapler. The patient is being treated with Imatinib Mesylate on an outpatient basis.

**DISCUSSION**

GIST is a descriptive term for intraabdominalmesenchymal tumours that arise mostly in the GI tract were definitely not carcinomas and failed to exhibit features of smooth muscle or nerve cells (MAZUR and CLARK ,1983) [4]. GIST cells bore
histopathological similarities with interstitial cells of Cajal (pacemaker cells). Both cells expressed KIT receptor tyrosine kinase (KIT RTK /CD117) [3]. Incidence of approximately 15% /million. A peak incidence in the fifth and sixth decades of life is noted and slightly more common among males [3, 4]. GISTs are most commonly located in the stomach and proximal small intestine, but can occur in any portion of the alimentary tract including the omentum, mesentery, and peritoneum, although, as reported here GIST may also have unusual origins from sites such as the pelvis (E-GIST) or Extra gastrointestinal stromal tumour [4].

Exact incidence of E-GIST not known because of rarity of reporting and cells of origin are controversial. Majority of E-GIST are >10 cm and histologically resemble interstitial GIST. As in present case tumour arising from pelvis (E-GIST) in a 60 year old male. Most patients with GIST present with symptoms of varying severity ranging from dysphagia, abdominal discomfort, altered bowel movements and bowel obstruction, to acute or chronic gastrointestinal hemorrhage and peritonitis due to perforation [3, 4]. In the present case the patient had a visible palpable tumor with significant weight loss and reported chronic constipation. Preoperative diagnosis of a GIST is uncommon due to their rarity varying presentation and the lack of distinguishing characteristics on imaging studies. The diagnosis of GIST is based on cellular morphology and immunophenotype. Histologically there are three types of GIST: Spindle cell (70%), epithelioid (20%), and mixed. Approximately 95% of GISTs carry an activating somatic mutation of CD117 (c-kit) [3]. Positive immunohistochemical staining for CD117 is a defining characteristic of GIST [3]. Tumor size and mitotic index are the two most important prognostic factors used for risk stratification of GISTs [5]. Outcomes in patients with GIST depend very much on clinical presentation and the histopathological features of the tumor. Thus the overall 5-year survival rate for all patients with GIST ranges from 28–60% but this range can be stratified based on disease progression. For patients presenting with localized primary disease the average survival rate is 5 years and those with metastatic or recurrent disease the average survival rate is approximately 10–20 months.

Contrast-enhanced CT scan is the first choice imaging modality for patients with suspected abdominal mass as it is useful for both preoperative staging and evaluation for metastatic disease [6]. There are no specific CT findings for GIST tumors although they typically appear as inhomogeneous masses with areas of necrosis and hemorrhage with viable tumor areas showing contrast enhancement [6]. FDG-PET use of 18F-fluorodeoxyglucose [18FDG] Positron emission tomography (PET) adds to functional imaging data [6]. Complete surgical resection is the treatment of choice and biological therapy with imatinibmesylate is recommended for cases with incomplete resection or unresectable tumors and when tumors have progressed to metastatic disease [1]. A more recent agent, sunitinib which inhibits vascular endothelial growth factor receptor (VEGFR) in addition to KIT and platelet-derived growth factor receptor alpha (PDGFRα) has proven to be effective in patients intolerant or refractory to imatinibmesylate [1].

CONCLUSION

Our case suggests possibility of GIST must be kept in mind while evaluating any case of abdominal lump. Occurrence of E-GIST is rare and treatment options are similar. The advent of molecularly targeted therapies revolutionized the management. Surgery remains the first choice of early stage and resectable disease. In female patients gynecologists should consider GIST in the differential diagnosis of patients presenting with an atypical ovarian mass.

REFERENCES


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