

Case Report

Extraintestinal Gastrointestinal Stromal Tumor arising from parietal wall: A rare entity

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Abstract

Extraintestinal gastrointestinal stromal tumors (E-GIST) are rare types of GIST most commonly involving the mesentery, omentum and retroperitoneum. Parietal wall E-GIST are very rare. Very few cases have been reported in the literature.

Keywords: GIST, parietal wall

Introduction

Extraintestinal GIST (E-GIST) are types of GIST that arise primarily outside the GI tract. Histologically and immunohistochemically they resemble their gastrointestinal counterpart¹. Approximately 80% are located in the omentum or mesentery, and the remainder develop in the retroperitoneum²⁻⁴. Very few cases of EGIST in the abdominal wall have been reported^{5,6}. Surgery is curative. Open surgery is indicated in large tumors to prevent risk of rupture. Laparoscopic surgery can be safely performed in small tumors (2-5cm). Prognostic factors in the case of EGISTs are not well described in the literature.

Case presentation

An 18yr old married female presented to the Tribhuvan University Teaching Hospital outpatient department in December 2014 with complaints of pain left upper abdomen on and off for 9 years. There was also complain of vague lump in left upper abdomen for 9 years. According to the patient the mass was gradually increasing in size. On examination there was a smooth, firm mass in left upper quadrant moving with respiration.

Ultrasonography of the abdomen showed 62 X 58 X 56mm well defined hypoechoic solid mass with cystic areas noted in left upper quadrant separate from solid organs. Contrast enhanced computed tomography of

the abdomen showed a heterogeneously enhancing well defined spherical soft tissue density mass in the left upper abdominal cavity likely to be arising from the omentum (Figure 1). Upper gastrointestinal endoscopy showed features suggestive of atrophic antral gastritis. Ultrasonography guided trucut biopsy was done. Immunohistochemistry staining showed tumor cells positive for CD117.

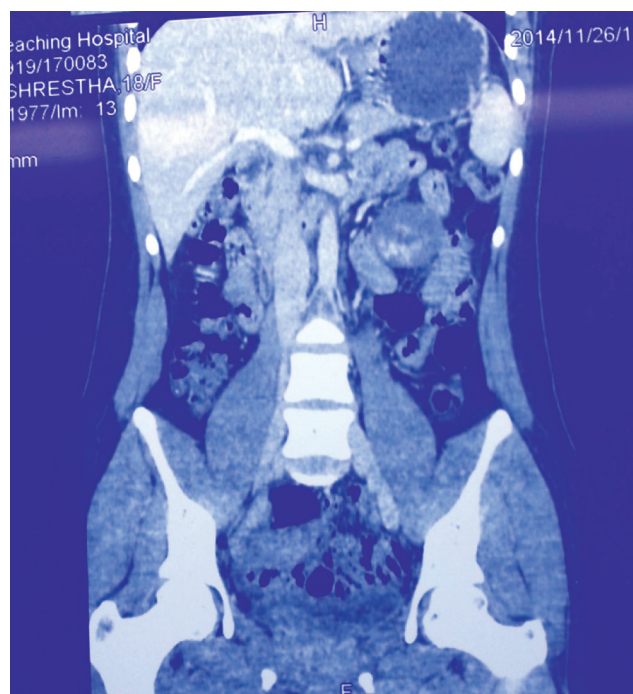


Figure 1

Blood investigations revealed a normal haemoglobin, normal counts, renal function test, prothrombin time.

Laparoscopic en bloc excision of the tumor was planned. Ten millimetres supraumbilical port was made and 5mm ports were made in the left iliac fossa and right hypochondriac region respectively (Figure 2).

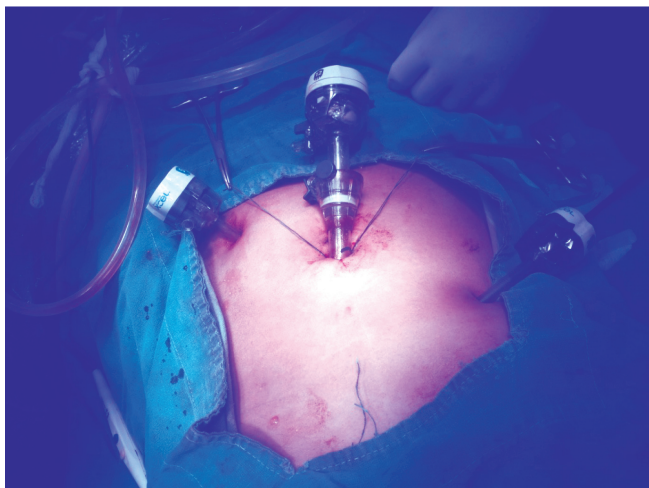


Figure 2

Surprisingly there was approximately 6 X 5 X 4 cm mass arising from the parietal wall of abdomen without any visceral attachment (Figure 3).



Figure 3

The mass was dissected from the parietal wall (Figure 4) and delivered through a midline incision in supraumbilical region (Figure 5). Other organs were normal. Cut section showed a solid mass with central cavity (Figure 6). Patient was discharged on post operative day two. Histopathology reports were consistent with spindle cell type of GIST.

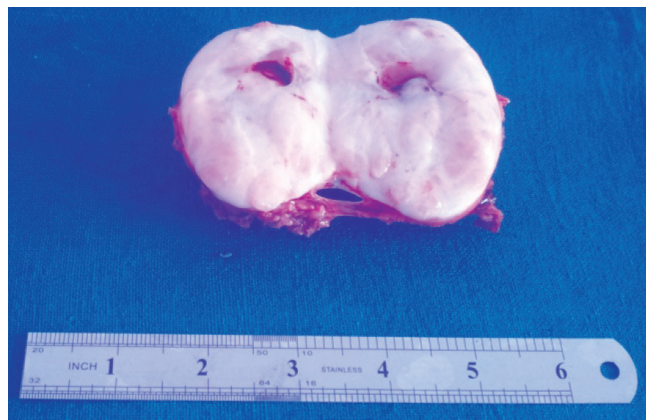


Figure 4

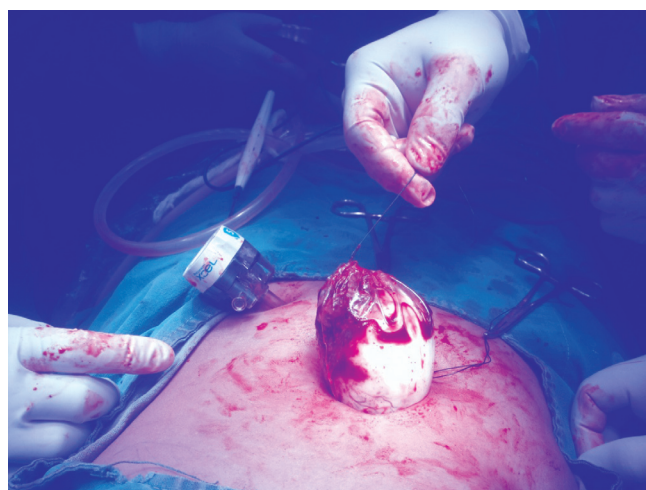


Figure 5

Discussion

Gastrointestinal Stromal Tumors (GISTs) are the most common mesenchymal tumors of the GI tract⁷ many of which are identified in the fifth or sixth decades, mainly occur in the stomach or small intestine, with a smaller percentage in other locations⁸. The pathogenesis is a gain of function mutations in the c-KIT gene which results in over expression of the KIT receptor also called CD117 or stem cell factor receptor. The counterpart that arises outside the gastrointestinal tract was termed as Extra gastrointestinal Stromal Tumor (EGIST) by Reith et al¹.

In our case immunohistochemistry showed CD117 positivity thus supporting a diagnosis of GIST and ruling out other soft tissue tumors. Intraoperative findings confirmed the origin of the tumor from the parietal wall.

A great percentage of EGISTs appear to be due to metastasis from a primary GIST⁸. In our case,

the diagnostic workups, including CT scans and endoscopies proved that the mass was likely to be a genuine EGIST.

Competing interests

1. The authors' would like to disclose that the interpretation of data or presentation of information in this manuscript hasn't been influenced by their personal or financial relationship with other people or organizations. 2. The authors haven't received reimbursements, fees, funding, or salary from an organization that may in any way gain or lose financially from the publication of the article, either now or in the future. 3. The authors aren't holding stocks or shares in an organization that may in any way gain or lose financially from the publication of the article, either now or in the future. 4. The authors aren't holding, or currently applying for, patents relating to the content of the manuscript. 5. The authors aren't receiving reimbursements, fees, funding, or salary from an organization that holds or has applied for patents relating to the content of the manuscript.

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Conclusion

E-GIST is a rare condition often confused with other soft tissue tumors. Early diagnosis requires a high index of suspicion in combination with different investigations.

Conflict of interest: None declared.

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