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Gastrointestinal Stromal Cell Tumor (GIST) Presenting as an Abdomino-pelvic Tumor

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Background: Gastrointestinal stromal tumors (GIST) are rare mesenchymal tumors that may mimic ovarian tumor on presurgical testing. These tumors are usually asymptomatic, often discovered accidentally during endoscopic or radiologic examinations,.

Case Presentations: In first case a 50 years old woman presented with irregular bleeding per vaginum, intermittent right lower abdominal pain and urinary frequency over the preceding 4 months. Physical examination and imaging studies revealed a pelvic mass of 18-week-gestational size. An exploratory laparotomy showed a huge fragile pelvic tumor, measuring 20×20 cm, arising from the jejunum. Immunohistochemical staining confirmed the presence of CD-117.

In second case an 53-year-old, postmenopausal, woman presented with abdominal fullness and constipation for the preceding 3-4 days. Physical examination and imaging studies revealed a huge pelvic mass, suggestive of a cystic degenerated myoma. An exploratory laparotomy revealed a large tumor originating from the ileum.

Conclusion: Gynecologists should keep in mind the possibility of non gynaecological tumors (GISTs) in the differential diagnosis in pelvic tumors.

Introduction

GISTs are mesenchymal tumors that represent 0.1-1% of all gastrointestinal malignancies¹. They occur most frequently in stomach (60-70%) followed by small intestine (25-35%), colon and rectum (5%) and oesophagus (<2%)². They are commonly asymptomatic and found incidentally during laparoscopy, surgical procedures or radiological studies. As regards their development, ~20-30% of GISTs show malignant behaviour³. Very rarely GISTs can mimic as gynaecological neoplasm⁴. Herein we report two cases of GISTs mimicking as gynaecological neoplasm on pre-surgical testing.

Case Presentation

Case 1

A 50 year old woman presented with irregular bleeding per vaginum, intermittent right lower abdominal pain and urinary frequency over the preceding 4 months. The general physical examination was negative, and a pelvic examination revealed an abdomino-pelvic mass of 18 week gestational size. Abdominal and transvaginal ultrasonography reported a $18.6 \times 20.4 \times 19.5$ cm irregular complex cystic solid mass arising from right adnexa. CT scan finding showed a large, well circumscribed heterogenous solid pelvic mass with a peripheral cystic component, which corresponds to the USG finding. CA-125 value was 130 U/ml (normal <35U/ml). The patient underwent surgery for a pelvic mass with suspected malignancy. Exploratory laparotomy showed a huge fragile pelvic tumor adhered to right adenexa, measuring 20×20 cm arising from jejunum. Uterus and left adenexa were normal. Pathology report revealed finding consistent with a GIST. Grossly the tumor was found to have arisen with in the muscularis propria of small bowel wall. Histologically tumor was composed of dense hypercellular infiltrate of spindle shaped cells.





Case 2.

A 53 year old, postmenopausal woman presented with abdominal fullness and constipation for the preceding 4 months. On pelvic examination an abdomino-pelvic mass of 14 week size was found. Uterus could not make out separately from the mass. USG revealed a pelvic mass 11×8 cm, suggestive of cystic degenerated myoma. An exploratory laparotomy revealed a large tumor originating from ileum. On histopathological examination, findings were consistent with GIST (Figure 1). Immunohistochemical staining confirmed presence of CD 117, Vimentin and desmin. Post operative period was uneventful.

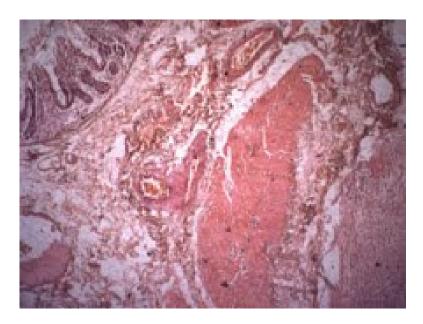


Figure 1. Histopathological Photograph Showing Gastrointestinal Stromal Cell tumor.

Discussion

GISTs are among the most common mesenchymal tumour of the gastrointestinal tract⁵. Almost 1/3rd of these are discovered incidentally during investigative or therapeutic procedures unrelated diseases⁶. Many of these tumours are found in 5th or 6th decade of life. The cell of origin of GIST is thought to be intestinal cells of Cajal (ICC), the pacemaker cells that controls GIST peristalsis⁷. The most common clinical presentation is gastrointestinal bleeding. Other signs include nausea, vomiting abdominal pain, weight loss, intestinal obstruction and abdominal distension. Few of these symptoms were experienced by the patient in our report⁸. There are few reports of GISTs presenting as pelvic mass, mimicking gynaecological neoplasms. In such patients, common different consideration includes pedunculated fibroid or ovarian neoplasm. In none of these patients was the correct diagnosis made pre-operatively⁴. The pre-operative diagnosis of GISTs is uncommon due to their rarity and different mode of presentation as well as the lack of distinguishing characteristics on imaging studies³. In our cases also diagnosis was made intra-operatively.

Diagnosis of GISTs solely based on USG can be difficult. They exhibit a variety of features, ranging from homogenous solid masses to heterogeneous solid masses with internal areas of hypoechogenicity corresponding to areas of necrosis and haemorrhage. Sometime due to large size of the masses, as in our cases, can make it difficult to find the origin on ultrasound. CT imaging plays an important role in identifying GISTs as well as assessing local spread and distant metastasis. A large mass with an exophytic



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growth pattern on CT is suggestive of a GIST⁴. Histologically, GISTs show different cellular patterns like spindle cells, epitheloid and rarely pleomorphic. The best defining feature is the presence of expression of c-kit (CD 117), a tyrosine kinase growth receptor and CD 34^{9,10}.

The main treatment has been surgical resection of the primary disease along with any metastasis to liver and peritoneal surface¹¹. Tyrosine kinase inhibitors such as imatinab and more recently, sunitinib have revolutionised the treatment of advance GISTs¹². Our reports suggest that we should keep in mind the possibility of a non-gynecological tumour among pelvic masses.

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