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Extra-gastrointestinal stromal tumor presenting as a huge peritoneal mass and mimicking as mesothelioma – A case report

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ABSTRACT

INTRODUCTION AND IMPORTANCE: Extra-gastrointestinal stromal tumors are group of soft tissue neoplasm, which originates outside the gastrointestinal tract comprising of less than 5% of the total gastrointestinal stromal tumors.

CASE PRESENTATION: A 67 years old male came with a history of vague abdominal pain, discomfort and loss of appetite. Per abdominal examination showed a palpable firm mass which was filling both the flanks. Radiological imaging revealed a large abdomino-pelvic mass with central necrotic areas. Exploratory laparotomy was done and the mass was excised intact from the sigmoid mesocolon. Histopathological diagnosis was given as extra-gastrointestinal stromal tumors.

CLINICAL DISCUSSION: Extra-gastrointestinal stromal tumor was first described by Miettinen et al. in 1999. The tumor can arise from the pleura, omentum, mesentery, retroperitoneum and prostate. The clinical presentation of the tumor depends on its location and the size of tumor. Patients with these tumors present with abdominal pain, followed by abdominal mass and distention. These tumors show pathological, immunohistochemical and molecular biological characters similar as that of gastrointestinal stromal tumor.

CONCLUSION: Extra-gastrointestinal stromal tumor is a rare tumor and can reach to a considerable large size before presenting with clinical symptoms especially if the tumor arises from the mesocolon. IHC study plays an important role to reach to the final diagnosis as the tumor can mimic mesothelioma in routine staining.

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1. Introduction

Gastrointestinal stromal tumors (GIST) are rare tumors of mesenchymal origin, accounting for less than 1% of the primary neoplasms of the digestive tract [1]. These tumors can affect any segment of the gastrointestinal tract from esophagus to anus, but can also occur in other locations in 10% of the cases, and, in such situations, these tumors are known as extra-gastrointestinal stromal tumors (E-GIST) [2]. E-GIST was first described by Miettinen et al. in 1999 [3]. These tumors show pathological, immunohistochemical and molecular biological characters similar to that of GIST [4].

We report a rare case of huge mesenteric extra-gastrointestinal stromal tumor presenting as a peritoneal mass. This case report has been reported in line with SCARE Criteria [5].

2. Patient information

2.1. Demographic details

A 67 years old male of Brahmin ethnicity from Sub-Himalayan valley of Pokhara in Nepal visited surgical outpatient department. He was a farmer by profession, belonged to low socio-economic status and was married for 40 years.

2.2. Patient presentation

He presented with vague abdominal pain, discomfort and sudden reduction of appetite for the past 2 months. Per abdominal examination revealed a palpable huge firm mass which filled both the flanks. General examination and other systems including respiratory and cardiovascular were within normal limits. His bowel and bladder habits were normal.

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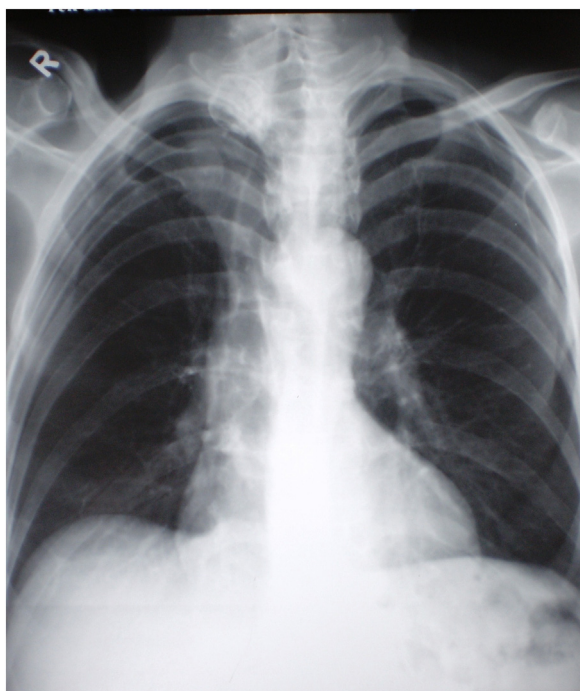


Fig. 1. Normal chest x-ray view of the patient.

2.3. Family and social history

There was no similar history of abdominal mass in his family and first degree relatives. He was non-smoker, non-alcoholic and there was no past history of exposure to chemicals or asbestos.

2.4. Laboratory and radiological findings

Routine hematological, serological, urine and coagulation tests were normal. Chest X-ray showed no pleural mass or bony lesion (Fig. 1). USG revealed a huge mass occupying the abdominal and pelvic cavity. Bilateral mild hydronephrosis was also noted. Contrast CT scan affirmed a large abdomino-pelvic mass (Fig. 2A) with central degenerative/necrotic areas (Fig. 2B). CT differentials of gastrointestinal stromal tumor or soft tissue sarcoma were given.

USG guided FNAC exhibited highly cellular smears composed of tumor cells arranged in clusters, sheets and scattered singly. The tumor cells were spindle shaped with moderate amount of cytoplasm and elongated nuclei (Fig. 3). Necrosis and atypical mitosis

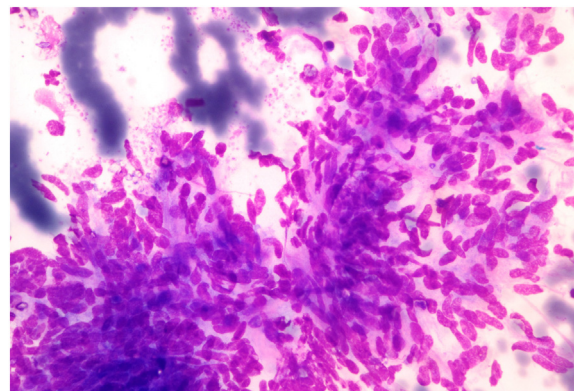


Fig. 3. Fine needle aspiration cytology picture showing clusters of spindle shaped cells. (H&E 400x).

were not noted in the aspirate. Cytology impression was given as spindle cell lesion and biopsy was advised.

2.5. Surgical intervention

Exploratory laparotomy was performed in a teaching hospital and the mass was excised intact from the sigmoid mesocolon. Operation was performed under general anesthesia by an experienced surgical team and the whole procedure took around 1 h. Patient was shifted to the post operative ward and was kept under antibiotic and analgesic coverage for 7 days. Abdominal stitches were removed and the patient was discharged on 7th post operative day. Patient was asked to follow up after 1 week.

Gross examination of the specimen revealed a huge mass measuring 28 × 23 × 16 cm (Fig. 4A). External surface was lobulated, dark to pale brown in color with presence of congested blood vessels. Cut section demonstrated lobulated, pale white solid mass with hemorrhagic and necrotic area measuring 14 × 12 cm (Fig. 4B).

On microscopic examination, the tumor composed of spindle shaped cells arranged in fascicles and bundles (Fig. 5A). The cells exhibited indistinct cytoplasmic membrane, pleomorphic elongated nuclei with inconspicuous nucleoli (Fig. 5B). Large areas of hemorrhagic necrosis were noted and atypical mitoses were 3/50 HPF. Routine Hematoxylin and Eosin (H&E) stain was misleading and the tumor was speculated to be a sarcomatous variant of mesothelioma. IHC study confirmed that the tumor cells were positive for CD117 (Fig. 6A), CD34 (Fig. 6B) and negative for S100 (Fig. 7A) and SMA (Fig. 7B). Hence, the final histopathological diagnosis of extra-gastrointestinal stromal tumor was confirmed.

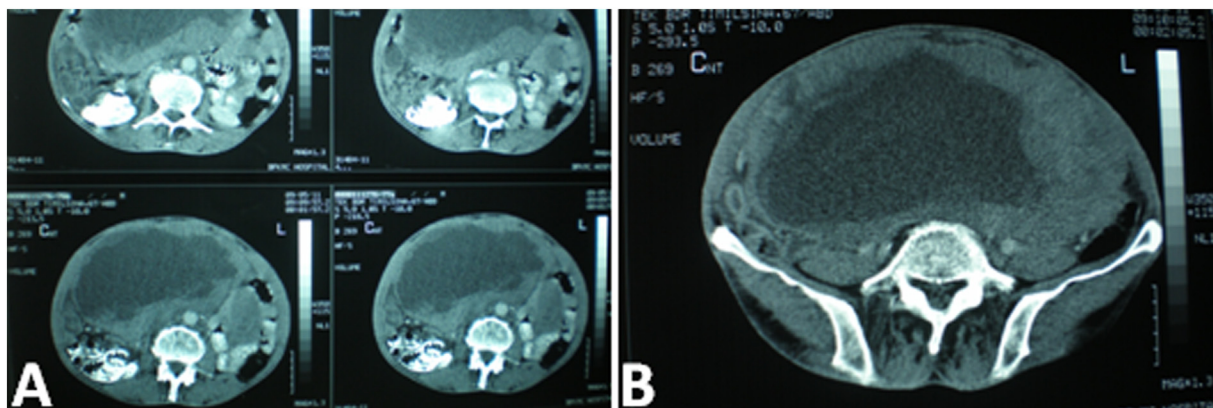


Fig. 2. Contrast CT abdomen, axial section displays large well defined heterogeneously enhancing necrotic mass occupying the abdomen displacing the viscera posteriorly (A). Contrast CT abdomen at lower level showing the mass extending into pelvis (B).

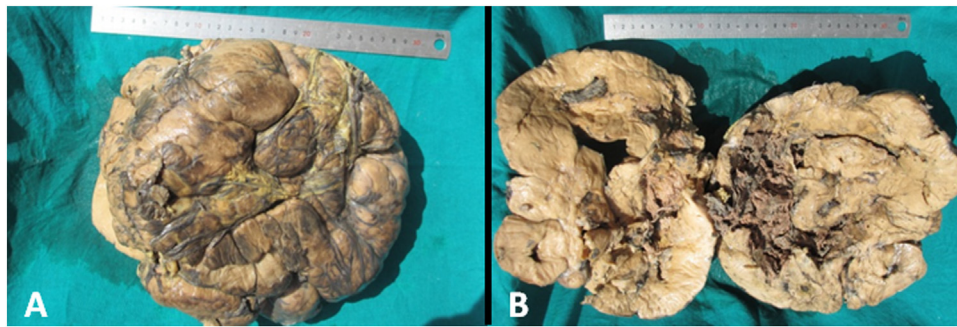


Fig. 4. Large dark to pale brown lobulated mass with congested blood vessels (A). Cut surface showed solid yellowish tumor with areas of necrosis and hemorrhage (B).

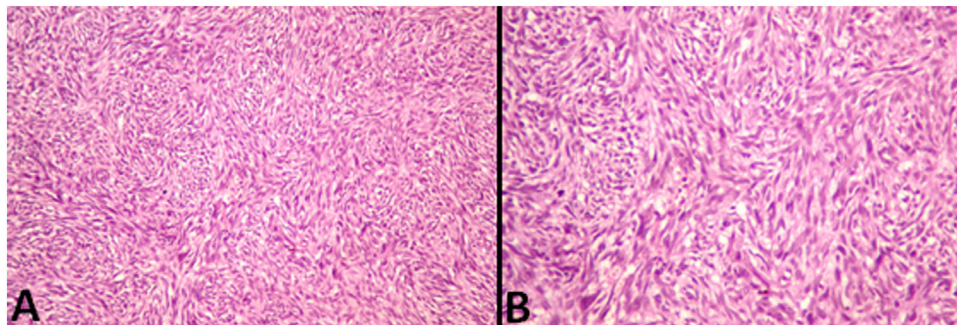


Fig. 5. Tumor showing sheets of spindle shaped cells in fascicles and bundles (A, H&E 200x). Tumor cells show scant cytoplasm and elongated nuclei. (B, H&E 400x).

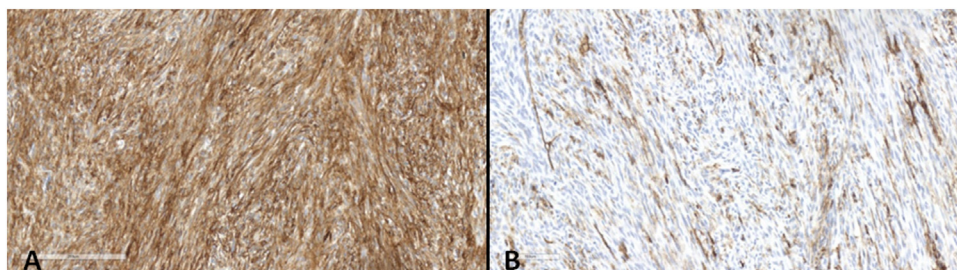


Fig. 6. IHC stain showed tumor cells positive for CD 117 (A) and CD 34 (B).

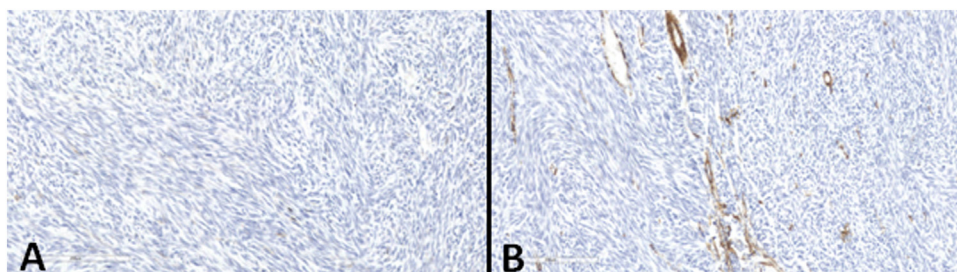


Fig. 7. IHC stain showed tumor cells negative for S100 (A) and SMA (B).

2.6. Follow-up and outcomes

The operative wound looked healthy on 14th post-operative day. There was no further complaint from the patient and he was referred to oncology department.

3. Discussion

Extra-gastrointestinal stromal tumor is defined as a gastrointestinal stromal tumor that occurs outside the gastrointestinal tract [1]. Although rare, GIST is the most common primary mesenchy-

mal neoplasm arising from the gastrointestinal tract and accounts for 1% of all gastrointestinal tumors. GISTs are identified in the fifth or sixth decades of life [6]. The main difference between GIST and E-GIST is the site of origin of the primary tumor, as GIST occurs throughout the gastrointestinal tract whereas E-GIST is a tumor without any connection with the intestinal wall [1]. Stomach is the most common site for the occurrence of GIST (50–60%), followed by the small intestine (20–30%), duodenum (5%), rectum (5%) and esophagus (1%) [4,7]. E-GIST can arise from the pleura, omentum, mesentery, retroperitoneum, liver, pancreas and prostate [4,8]. Studies have shown that the most common anatomic location for

the E-GIST is intra-abdominal cavity, including mesentery or omentum while retroperitoneum is the second most common site of origin. [5,9,10] In the present study, the tumor was present in the intra-abdominal cavity which was arising from the mesocolon of sigmoid colon.

The clinical presentation of GIST broadly depends on tumor location and the size of tumor. For mesenteric E-GIST, tumor appears to have enough space to grow and may show clinical symptoms after a significant period of time when the tumor has reached a considerable size [11]. Patients with E-GIST presents with abdominal pain, followed by abdominal mass and distention. Unlike gastrointestinal GIST, bleeding is rarely seen in E-GIST [4]. In our case also, the patient came for consultation with a complaint of vague abdominal pain when the tumor has reached to a huge size of 28 cm.

The omental E-GIST may more closely resemble gastric stromal tumors whereas the mesenteric tumors may mirror small bowel stromal tumors [9]. Studies have revealed that E-GIST tumors show positive immunohistochemistry in 93.3% for CD117 (c-kit receptor), 70% for CD34, 44% for neuron-specific enolase, 26% for smooth muscle actin, 10% for S100 and 4% for desmin [2,9]. However, there are GISTs that have a mutation of platelet-derived growth factor receptor alpha (PDGFRA) instead of c-KIT and therefore they do not show the characteristic CD117 positive immunostaining [6]. The mutation analysis of c-KIT and PDGFRA is useful for confirming CD117 negative GIST [7]. Thalheimer et al. reported an E-GIST of abdominal wall in a 40 years old patient with 24 cm sized tumor with low mitotic index (1/50 HPFs), in which tumor cells showed positivity for CD117, CD99, CD34, vimentin, and actin [12]. In the present case, the tumor cells were positive for CD117 and CD34 but negative for S100 and SMA.

Mitotic figures are considered as the single most important microscopic feature for considering the tumor as malignant and can be taken as a powerful prognostic indicator. Various studies have shown that if the mitotic figures are more than 5/50 HPF, then the outcome is unfavourable for GIST [13–16]. Reith JD et al. in their studies have reported that high cellularity, mitotic index more than 2/50 HPF and presence of necrosis are indicative of aggressive clinical course for E-GIST [9]. The present case showed areas of tumor necrosis and mitosis 3/50 HPF.

GIST needs to be distinguished from leiomyoma, schwannoma, inflammatory fibroid polyps and fibromatosis [2,7]. Peritoneal mesotheliomas with sarcomatoid differentiation may mimic E-GIST and needs to be differentiated [17]. In our case also, the tumor cells resembled sarcomatoid mesothelioma in routine H&E stain. E-GIST and other mesenchymal tumors can be diagnosed by a combination of routine histopathological examination and immunohistochemical staining [2,7]. The majority of stromal tumors stain positively with CD34, a marker of myeloid progenitor cells, endothelial cells, and some mesenchymal lesions, which help to separate GIST from leiomyomas and schwannomas which are CD34 negative [18]. Though mesothelioma may resemble E-GIST in H&E stain, they are positive with calretinin, WT-1, cytokeratin 5/6, and D2–40 but are negative with CD 117 and CD34 [17].

The treatment of choice for the E-GIST is the en bloc resection of tumor with macroscopically negative margin followed by adjuvant imatinib as most of the patients fall into high-risk category for recurrence [4]. In borderline resectable primary tumors, preoperative imatinib is a possible option but there are no randomized trials assessing the benefit of neoadjuvant treatment [19]. Tumor location is also one prognostic factor for GIST, and it was considered that E-GISTs are more aggressive than gastric GISTs [11].

4. Conclusion

Extra-gastrointestinal stromal tumors are rare tumors of mesenchymal origin arising outside the gastrointestinal tract which should be differentiated from sarcomatous variant of mesothelioma and various other mesenchymal tumors. In the present context, IHC study played an important role as cellular differentiation was not conclusive only on light microscopy.

Declaration of Competing Interest

Authors do not have any conflicts of interest.

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Ethical approval

Ethical approval for the case report study is not required.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

1. Dilasma Ghartimagar – Study concept, data collection and paper writing.
2. Arnab Ghosh – Study design and data analysis.
3. Manish Kiran Shrestha – Data interpretation, editing paper writing.
4. Hemant Batajoo – Study design and paper editing.
5. OP Talwar – Data analysis and editing paper writing.

Registration of research studies

Not applicable.

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Provenance and peer review

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