Ruptured primary extra-gastrointestinal stromal tumour of the greater omentum mimicking a liver hemangioma

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Abstract

Background
Gastrointestinal stromal tumours (GISTs) represent the majority of primary non-epithelial neoplasms of the digestive tract, most frequently expressing the KIT protein detected by the immunohistochemical staining for the CD117 antigen. Extra-gastrointestinal stromal tumors (EGISTs) are neoplasms with overlapping immunohistological features, occurring in the abdomen outside the gastrointestinal tract with no connection to the gastric or intestinal wall.

Case Presentation
We here report the clinical, macroscopic and immunohistological features of an EGIST arising in the greater omentum of a 74 year-old man.

Conclusion
The EGISTs in the greater omentum can grow slowly in the abdomen for a long time without clinical appearance. In most cases a preoperative diagnosis is not possible, and the patient undergoes a surgical operation for the generic diagnosis of “abdominal mass”. EGISTs being highly vascular tumours can show enhancement findings mimicking hemangiomas on computed tomography. Surgery remains the mainstay of treatment in non-metastatic EGISTs.

Introduction
Gastrointestinal stromal tumors (GIST) are uncommon mesenchymal spindle-cell or epithelioid neoplasms, located mainly with higher frequency in the stomach and small bowel. Extra-gastrointestinal stromal tumors tend to present in fewer than 5% of cases; and tend to be more common in patients over the age of 50 years. They originate primarily from the mesentery, omentum or peritoneum. They are characterized by the expression of KIT (CD117), a transmembrane tyrosine kinase receptor for stem cell factor. Their presumed cell of origin is the interstitial cell or epithelioid neoplasms, occurring in the abdomen outside the gastrointestinal tract with no connection to the gastric or intestinal wall.

Case Report
A 74 year old man presented to us with an incidentally detected abdominal mass on routine ultrasonography done during a master health checkup in September 2012. The mass was in close proximity to the left lobe of liver and an abdominal computed tomography demonstrated a 5 x 4 x 4 cms mass lesion with enhancement suggestive of a liver hemangioma. The patient refused surgery for the mass and was lost to follow up. He presented again in February 2013 with complaints of increase in the size of the abdominal mass, weight loss and early satiety. Abdominal CECT was repeated which demonstrated a rapid growth in the size of the mass to 11 x 7 x 6 cms. (Figure 2) In view of the large size and enhancement features on abdominal CECT it was decided to do an abdominal angiography and attempt angioembolization of major feeding vessels to the tumour. However abdominal angiogram did not reveal any abnormal feeding vessel that could be embolised. A diagnosis of a liver hemangioma or gastric GIST was made. Patient underwent a diagnostic laparoscopy followed by a midline laparotomy. There was moderate ascites. A large mass was found to be arising from the greater omentum. The mass was found to be supplied by the omental vessels. There was no connection between the stomach wall and the mass except for a small serosal adhesion. There were peritoneal nodules identified suggesting an intraperitoneal rupture of the mass. The mass was excised completely along with the omentum, and the peritoneal nodule. Grossly, an irregular lobulated gray white mass measuring, 17 × 14.5 × 8.5 cm in size, showed a breach measuring 7 x 3.5 cm with attached omentum measuring 30 x 5.5 x 1 cm. The cut surfaces were lobulated, convoluted and showed congestion, cystic change and foci of calcification. On sectioning the omentum, multiple nodules were noted. Histologically the tumour showed portions of a neoplasm composed of sheets of monotonous population of spindle shaped cells with indistinct cell borders, oval vesicular nucleus and prominent nucleoli. (Figure 1) Focal lobular pattern was noted. Also noted were small capillary sized blood vessels with angiocentric arrangement of tumor cells. The mitotic rate was 2-4/50hp. The peritoneal nodule also showed similar neoplastic features. Immunohistochemistry markers done were CD117, CD34, EMA, S100, SMA, Ki67, Desmin. The tumor cells were positive for CD117 (2 to 3+, 30%- 40%) and CD34 (3+, 80%). The Ki67 index was 8-10%. The rest of the markers were negative. These findings strongly supported a diagnosis of low risk EGIST of the greater omentum. The patient had an uneventful postoperative course. He was treated by administration of Imatinib mesylate 400

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mg/day as an adjuvant postoperative molecular targeting chemotherapy and has been living disease-free for 9 months now. This work conforms to the values laid down in the Declaration of Helsinki (1964). The protocol of this study has been approved by the relevant ethical committee related to our institution in which it was performed. All subjects gave full informed consent to participate in this study.

Discussion
EGISTs although arising outside the gastrointestinal tract, share histological features with their gastrointestinal counterpart. The data regarding clinical, pathological and prognostic features of EGISTs are very few. EGISTs are rare tumors. Todoroki et al. described one case citing 28 cases previously reported in the English-language literature.

The prognostic factors indicating the malignant potential of GISTs include mitotic rate, tumor size and location. Predicting the potential biological behavior of the omental EGISTs remains difficult. Miettinen et al. examined nine cases of omental EGISTs and seven cases of mesenteric EGISTs. Omental EGISTs seemed to have a more favorable behavior, typically showing low mitotic counts, whereas mesenteric EGISTs appeared more aggressive (higher mitotic activity, frequent malignant behavior). No tumor-related deaths were documented during the follow-up in the nine patients with omental EGIST.

Yamamoto et al. examined the clinico-pathological features, prognostic factors, c-kit and PDGFRA mutations in 39 cases of EGISTs including three omental tumors. These authors have defined three categories on the basis of a combination of the mitotic rate and MIB-1 labelling index: the high-risk group (>or=5/50 HPF with >or=10% Ki-67), the intermediate-risk group (>or=5/50 HPF with <10% Ki-67, or, <5/50 HPF with >or=10% Ki-67), and the low-risk group (<5/50 HPF with <10% Ki-67). Surgery remains the standard treatment for non-metastatic EGIST in the greater omentum.

The accurate radiological follow-up (abdominal CT) has been considered the approach of choice in the control of the disease.

Conclusion
The EGISTs in the greater omentum can grow slowly in the abdomen for a long time without clinical appearance. In most cases a preoperative diagnosis is not possible, and the patient undergoes a surgical operation for the generic diagnosis of "abdominal mass". EGISTs being highly vascular tumours can show enhancement findings mimicking hemangiommas on computed tomography. Surgery remains the mainstay of treatment in non-metastatic EGISTs.

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