

A Mobile Extra-Gastrointestinal Stromal Tumor

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Abstract

A 65-year-old man presented with epigastric pain. A CT scan carried out at admission demonstrated a large 10.5 cm mass in the right lower abdomen, which moved to the left upper abdomen on a MRI scan carried out 11 days later. The patient had a negative diagnostic laparotomy one year earlier for the same symptoms. He was initially planned for a percutaneous biopsy of the mass, however, had an emergency laparotomy due to massive hematochezia which found a large mesenteric tumor adherent to the proximal jejunum. Histopathology results confirmed an extra-gastrointestinal stromal tumor in the mesentery of the small intestine.

Keywords

Extra-gastrointestinal stromal tumor, Mesentery, Abdomen, Mobile

Abbreviations

MRI: Magnetic Resonance Imaging; CT: Computed Tomography; ¹⁸F-FDG: 18-Fluorine-fluorodeoxyglucose; PET: Positron Emission Tomography; GIST: Gastrointestinal Stromal Tumor; EGIST: Extra-Gastrointestinal Stromal Tumor; GRE: Gradient-Recalled Echo

Case

A 65-year-old man was referred for investigation of upper abdominal pain, on a background of having a negative diagnostic laparotomy a year prior for similar symptoms. The initial CT demonstrated a large mass in the right lower abdomen measuring 8.5 cm x 10.5 cm x 10.5 cm (Figure 1). Due to the previous negative diagnostic laparotomy, a joint decision was made with the patient for further investigations to be undertaken to help guide surgical management. Interestingly, an MRI which was carried out 11 days later demonstrated that the mass had migrated from the right lower to left upper abdomen, with features highly suggestive of a large mobile mesenteric malignancy (Figure 1). The patient subsequently had a gastroscopy and was further investigated with a Gastrografin small bowel series, the results of which were unremarkable.

Based on the results of the CT and MRI which raised high suspicion for a malignant process, an ¹⁸F-FDG PET/CT study was performed to further characterize and stage the lesion, to guide further management. The ¹⁸F-FDG PET/CT study demonstrated moderate FDG uptake in the mass (Figure 1), in keeping with an FDG avid malignancy, with no evidence of FDG avid

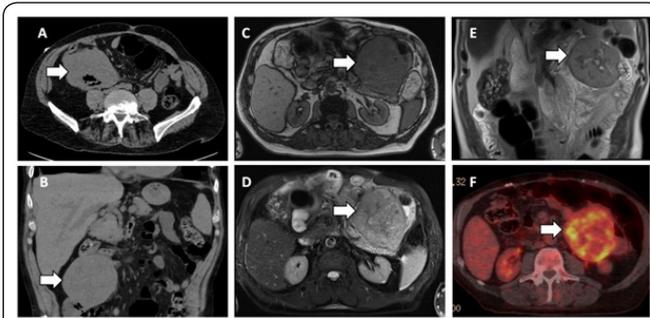


Figure 1: Non-contrast axial CT (A) and coronal CT (B) images demonstrate a large soft tissue mass in the right lower abdomen measuring 8.5 cm x 10.5 cm x 10.5 cm which is predominantly solid with locules of gas (arrows). Axial out-of-phase T1-weighted GRE sequence MRI (C), axial T2-weighted MRI (D) and coronal T2-weighted MRI (E) images demonstrate translocation of the same large mass to the left upper abdomen, which is hypointense on T1 and hyperintense on T2 with irregular enhancement (arrows). Axial fused FDG PET/CT (F) image demonstrates moderate FDG uptake in the lesion (arrow).

metastasis. The combined CT, MRI and ^{18}F -FDG PET/CT findings were compelling enough for consideration of a repeat laparotomy with an aim to excise the mass, however, histopathological confirmation prior to surgical management was favored. Unfortunately, while awaiting a percutaneous biopsy of the mass, the patient had an episode of massive hematochezia. This led to an emergency laparotomy which found a large mesenteric tumor adherent to the proximal jejunum. Histopathology results confirmed an extra-gastrointestinal stromal tumor in the mesentery of the small intestine (Figure 2). Post-operatively, the patient was admitted to the intensive care unit, stepped down to ward based care after 2 days and was successfully discharged from hospital 1 week later. On follow up in the surgical outpatient clinic in 1 month, he was well, symptom-free and reported complete resolution of his previous upper abdominal pain.

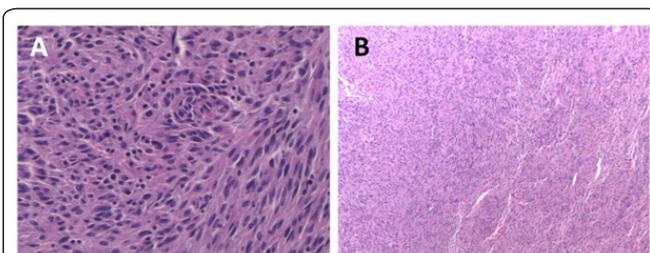


Figure 2: Histopathology slides (A, B) from the surgical resection confirm an extra-gastrointestinal stromal tumor (spindle cell type) in the mesentery of the small intestine, demonstrating compact sheets of spindle cells with focal areas of palisading pattern. Patchy areas of hemorrhage are demonstrated with no definitive necrosis.

Discussion

Extra-gastrointestinal stromal tumors (EGISTs) of the mesentery are rare, with case studies scattered throughout the literature [1, 2]. Gastrointestinal stromal tumors (GISTs) are mesenchymal neoplasms which affect the gastrointestinal tract, most commonly occurring in the stomach and proximal small intestine, only rarely occurring outside the gastrointestinal tract (where they are known as EGISTs), in places such as the

mesentery [3]. While GISTs themselves are rare and estimated by one study to have an annual incidence rate of 0.78 / 100000 [4], only less than 5% of these GISTs are EGISTs [5]. GISTs primarily occur in the older and middle-aged population, with a median age of diagnosis at 64 years old. It also demonstrates a predilection for males compared to females, occurring 36% more commonly in men.

The most common anatomic distribution of GISTs is the stomach (55%), followed by the small intestine (28%), colon (2.9%) and rectum (2.7%) [4]. The clinical presentation of GISTs and EGISTs can be quite varied, depending on the location of the primary tumor. Most commonly, patients present with occult or overt gastrointestinal bleeding (54%), followed by pain and upper abdominal discomfort (17%), while a small proportion of patients may remain asymptomatic (4%) [6]. It is thought that GISTs arise from CD34-positive interstitial cells of Cajal, which can be considered as the pacemaker cells of the gastrointestinal tract [7, 8]. EGISTs, which occur outside the gastrointestinal tract in sites such as the omentum, peritoneum and retroperitoneum, are believed to arise from interstitial cells of Cajal which were dispersed out of the gastrointestinal tract during embryogenesis [9].

The histopathological appearance of GISTs fall into 3 categories, which are the spindle cell (70%), epithelioid (20%) and mixed (10%) types; the cells also usually demonstrate immunopositivity to KIT (CD117) immunohistochemical staining [7]. Although their biologic behavior may vary, all GISTs are potentially malignant and thus, stratifying risk is more suitable rather than definitely categorizing them as benign or malignant [10]. Generally, large tumors (greater than 5 cm in size) which are lobulated, exophytic, heterogeneously enhancing and demonstrate evidence of regional involvement on imaging, high mitotic index, primary location outside the stomach (EGISTs in particular) and tumor rupture are considered to be poor prognostic factors, which are associated with higher metastatic potential and higher rates of relapse [11-13].

The mainstay of treatment for operable GISTs is surgical resection, with consideration of adjuvant tyrosine kinase inhibitor therapy (such as imatinib) for high risk patients [14]. While reported survival times in the literature vary depending on the age of the study, with the advent of systemic therapy, excellent 5-year rates of up to 90% recurrence-free survival and 95% overall survival have been recorded [15, 16].

The patient's CT, MRI and PET images were consistent with the typical findings of an EGIST, demonstrated as a large soft tissue density mass with central necrosis on CT, hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging on MRI, and FDG avid on PET [17-19]. Differential diagnoses of a mesenteric mass include other primary mesenteric tumors (such as desmoid or carcinoid tumors, which are also rare), secondary metastasis from other cancers and lymphoma [20].

While it has been postulated that mesenteric EGISTs have a certain degree of mobility owing to flexibility of the mesentery [21], to our knowledge, our case report is the

first to demonstrate and confirm movement of a mesenteric EGIST on serial imaging to such a large extent. This could have been a major contributing factor in our patient's previous negative diagnostic laparotomy, owing to difficulty in locating the tumor. Other possible reasons could be that the mass was previously smaller at the time of the initial laparotomy or that the surgical exploratory technique did not fully account for this mobility of the tumor. Therefore, clinicians should be aware that mesenteric EGISTs can be particularly mobile and take this into consideration when planning an approach for surgical exploration.

Conflict of Interest

The authors declare no conflict of interest.

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