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Case Report

# EXTRAGASTROINTESTINAL STROMAL TUMOR WITH RETROPERITONEAL LOCATION: A RARE CASE REPORT

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**Abstract**

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tissue-derived tumors of the gastrointestinal tract. They occur rarely outside the digestive tract, and they are defined as extragastrointestinal stromal tumors (EGISTs). As it is the case for GISTs, the cases with inoperable EGIST may also respond dramatically to the treatment with oral imatinib mesylate. Herein, we reported a 70-year-old patient admitted with a giant retroperitoneal mass, diagnosed with inoperable EGIST and responded dramatically to imatinib treatment.

**Keywords: Extragastrointestinal stromal tumor; Imatinib mesylate; Retroperitoneal location**

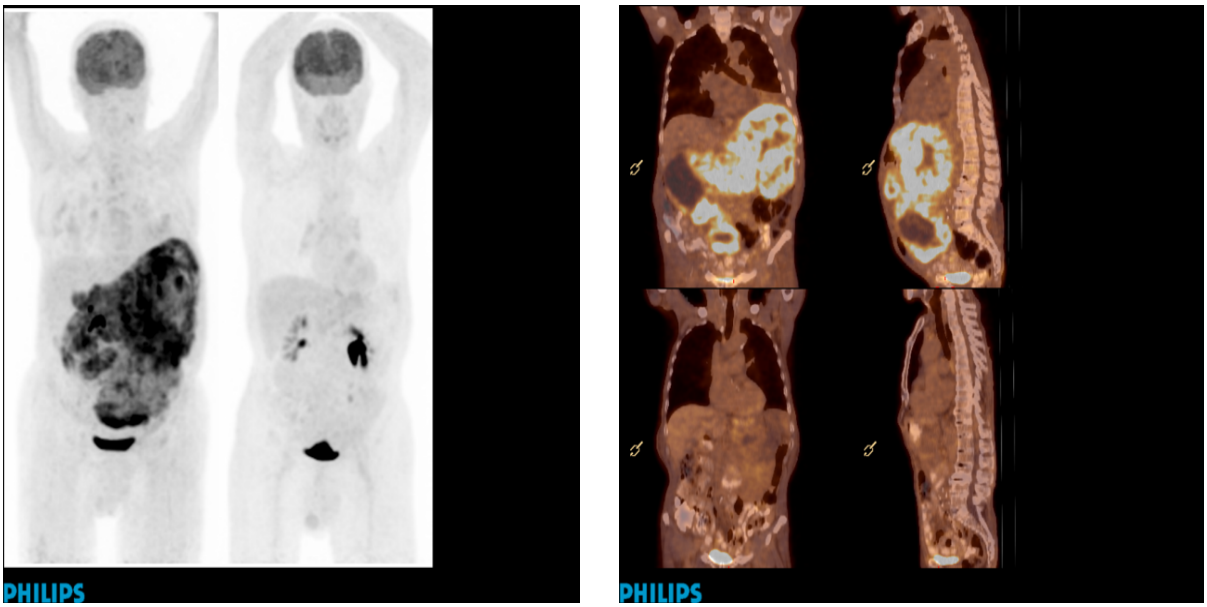
## Introduction

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tissue-derived tumors of the gastrointestinal tract and originate from the cajal cells in the gastrointestinal tract after the mutation of some tyrosine kinase receptors, such as c-kit and platelet-derived growth factor receptor alpha oncogene (PDGFRA). (1) They are mostly localized to the stomach and jejunum (60-70%). They are less frequently encountered in small intestine (20-30%), colon-rectum (5-12%) and esophagus (2-5%) in the gastrointestinal tract .(2) Less than 10% of GISTs are located outside of the digestive tract (omentum, mesentery, retroperitoneum, liver), and they are defined as extragastrointestinal stromal tumors (EGISTs). (3) EGISTs are also similar to GISTs in terms of histologic appearance and immunophenotypic profile but they show a more aggressive clinical course.

## Case Report

A giant retroperitoneal mass lesion was detected in the abdominal CT conducted by the general surgery clinic to which the 70-year-old male patient was admitted with the complaints of swelling and pain in the abdomen for three months. Laparotomy was performed in the surgical clinic. The case was considered inoperable and a biopsy was taken for diagnostic purposes. Histopathological examination revealed that the mass was a c-kit (CD117) and CD34 positive extragastrointestinal stromal tumor with high mitotic index. No

structure belonging to gastrointestinal system components was observed in the histopathologic examination. The Positron Emission Tomography (PET / CT) performed on the patient who was evaluated in our clinic demonstrated a giant mass lesion with lobulated contour measuring approximately 26 x 17 x 37 cm, starting from the basal part of the lower lobe of the left lung and adjacent to the left ventricle, filling the abdomen, extending till the pelvis, displacing the intestines, left kidney and main vascular structures, including wide hypometabolic and necrotic areas and showing intense 18-fluoro-deoxyglucose (FDG) uptake (SUVmax: 14.6). Oral imatinib mesylate treatment was initiated in the patient who had good performance status. Marked regression (15 × 11 × 9 cm, SUV max: 3.5) and clinically dramatic response were observed in the PET / CT of the patient that was assessed at the third month of treatment. The patient who is under imatinib treatment is still being followed up in our clinic. (Figure 1, 2)




**Figures 1, 2:** Pre-treatment and post-treatment FDG-PET/CT images of our patient

**Discussion**

GISTs originate from the Cajal cells in the gastrointestinal tract after the mutation of some tyrosine kinase receptors such as c-kit and platelet-derived growth factor receptor alpha oncogene (PDGFRA). EGISTs are derived from the multipotent mesenchymal stem cells which are the precursors of Cajal cells. Oncogenic c-kit gene mutations occur most commonly in exon 11 and to a lesser extent in exon 9. One of the important features defining GISTs is that the expression of CD117 (C-kit protein) is observed in almost all of these tumors. GISTs exhibit immune-histochemical staining characteristics which facilitate diagnosis The positivity rate of C-kit (CD117) is 95%, of CD34 is 60-70% and of smooth muscle actin is 30-40% .(4) A similar immunohistochemical staining pattern is observed in EGISTs. EGISTs frequently occur in the momentum, mesentery and retroperitoneal areas apart from the gastrointestinal tract. (3) However, atypical localizations such as pancreas, liver, gallbladder, bladder, pleura, prostate, seminal vesicle and vena cava inferior were also reported in the literature. (5-13) Retroperitoneal liposarcoma, soft tissue sarcomas, lymphoma and germ cell tumors should be considered in the differential diagnosis of the patients presenting with a giant retroperitoneal mass as in our case. The basic clinical findings of retroperitoneal EGISTs are mass and pain in the abdomen.

Also, nausea, vomiting and weight loss may be observed.

As for GISTs, the most important prognostic factors determining survival and relapse in EGISTs are the size and mitotic rate of the tumor. The most effective treatment of EGISTs is surgery as it is the case for GISTs. Their response to conventional chemotherapy agents is very low and they are radioresistant tumors. Imatinib mesylate (400 mg/day orally), which is a c-kit receptor and specific inhibitor of tyrosine kinase, is the first choice treatment for inoperable cases, cases with relapse and metastasis or cases whose general condition is not suitable for surgery and it changes the prognosis of patients dramatically as in our case. (14) For efficacy, the drug should be used for long-term (at least three months).

**Conclusion**

While the clinical, pathological and prognostic characteristics of GISTs are better understood today, EGISTs are less common and their biological behaviors are not known exactly. As the case reports, like our case, with dramatic response to imatinib mesylate therapy in a short period of time increase, the results of long-term follow-up of these cases will provide a better understanding of the biological behaviors of these tumors.

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