



## A rare solid tumor of the retroperitoneum with venous extension and lung metastasis: Extra-gastrointestinal stromal tumor

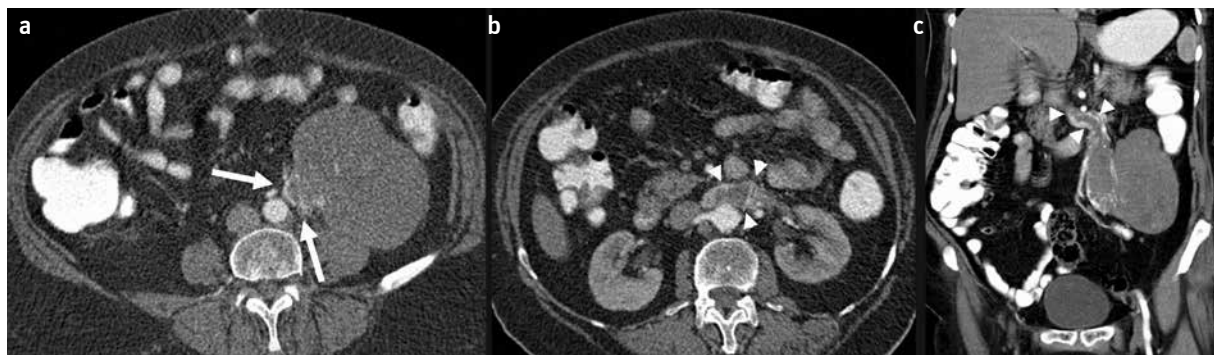
To the Editor,

We read with great interest the paper published by Al-Jehani et al. (1) in the Turkish Journal of Gastroenterology along with the other related papers regarding extra-gastrointestinal stromal tumors (E-GIST).

Retroperitoneal E-GIST is a very rare tumor and a total of 58 cases have been reported in the literature (2). The distinction of E-GIST from gastrointestinal stromal tumors (GIST) is made by proving the origin outside the alimentary system and ruling out a concurrent neoplasm in the gastrointestinal tract. In this letter, we aimed to report a very rare form of E-GIST arising from the retroperitoneal region, demonstrating lung metastasis and atypical feature such as venous extension.

A 72-year-old woman with complaints of left lower quadrant fullness and palpable mass that started 1 year before was admitted to the surgery department of our institution. On physical examination, a smooth contoured, hard mass without tenderness was detected in the left lower quadrant. Laboratory examination and tumor marker levels were unremarkable. Following physical examination and blood tests, the patient

was referred to computed tomography (CT). Imaging revealed a solid mass in the left retroperitoneal region measuring 10x10x12 cm anteroposteriorly, mediolaterally, and craniocaudally, respectively. The mass lesion demonstrated heterogeneous predominantly peripheral contrast enhancement with left renal vein extension via the left gonadal vein (Figure 1). Metastatic nodules in the lung were seen in thoracic images (Figure 2). Owing to the retroperitoneal localization and venous extension, the preliminary diagnosis of leiomyosarcoma was made, and following imaging, an ultrasound-guided Tru-cut biopsy was performed. The diagnosis of E-GIST was made after pathological evaluation with hematoxylin-eosin and immunohistochemical staining (Figure 3). Malignancies arising from the female reproductive tract and other possible concurrent neoplasm in the gastrointestinal system were ruled out using positron emission tomography (PET)/CT before treatment. The patient was initially managed using effective target therapy with an oral tyrosine kinase inhibitor imatinib mesylate for lung metastases. The patient remained stable for an interval following imatinib mesylate treatment. However, a progression was observed in the primary tumor and lung metastases despite 3 months of therapy. The medication was replaced with

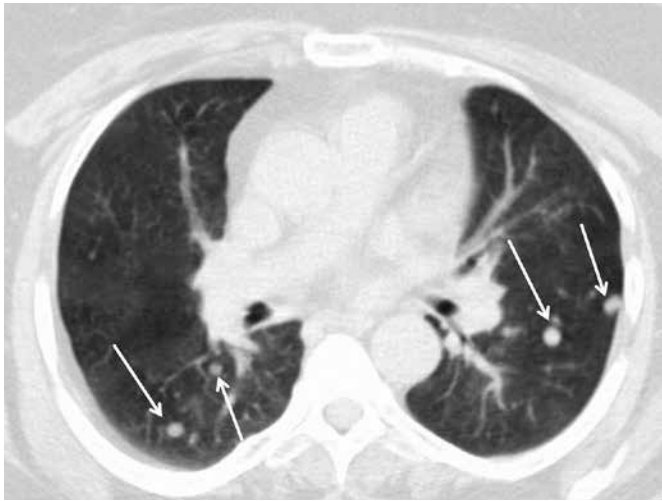


**Figure 1. a-c.** Axial contrast enhanced (a,b) and coronal reconstruction computed tomography (CT) images (c) show hypoattenuated, well-defined solid mass lesion in the retroperitoneal region. Peripheral enhancement is seen in the medial portion (arrows). Note that the descending colon is displaced by the mass anteriorly. Left renal and gonadal venous extensions are seen (arrowheads).

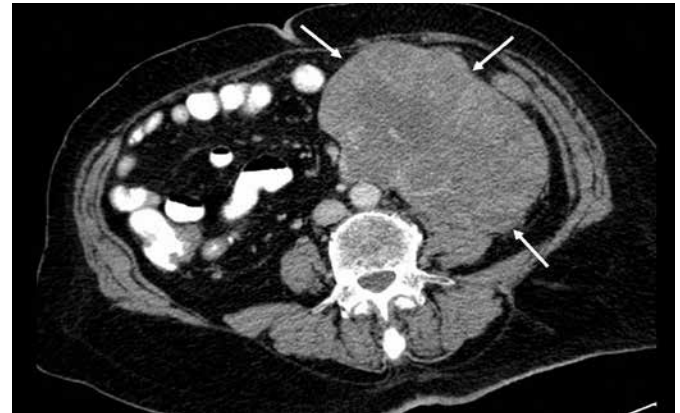
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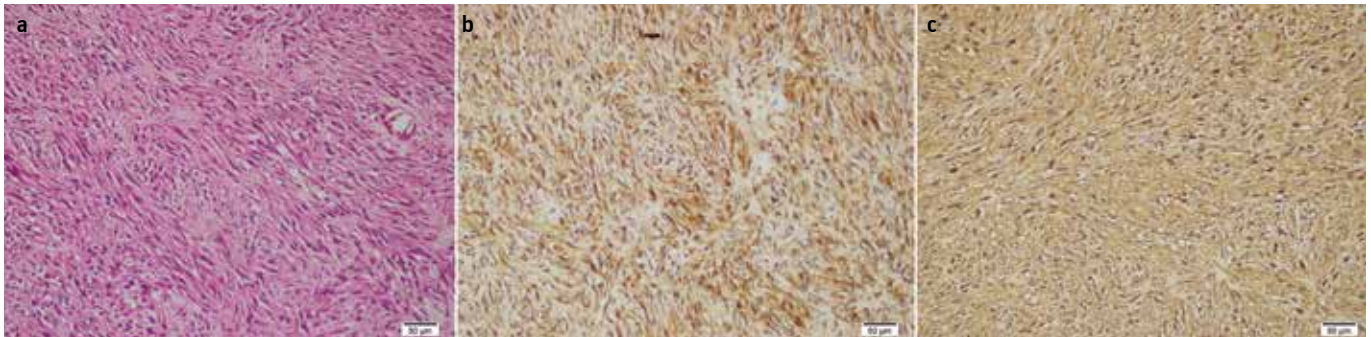
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**Figure 2.** Axial thoracic CT image demonstrates metastatic lung nodules (arrows).



**Figure 4.** Axial contrast enhanced CT image after 3 months of medical treatment with imatinib mesylate shows enlargement of E-GIST. The mass is 12.5x13.5x17.5 cm in size (arrows). Note that the diameters of the tumor have increased significantly compared with the first CT scan which was 10x10x12 cm in size.



**Figure 3. a-c.** Histopathologic views include hematoxylin and eosin stain of the extra-gastrointestinal stromal tumor (E-GIST) from the retroperitoneal region (20x) (a), CD-117 (c-KIT) stain of the E-GIST with positivity (20x) (b) and CD-34 stain of the E-GIST with positivity (20x) (c). According to the pathology report, smooth muscle actin, desmin, and S-100 protein are positive with a value greater than 50%.

sunitinib malate owing to the progression (Figure 4). To assess the response to medical treatment, the patient is still followed at short intervals.

Vascular invasion and spreading throughout the vessel trace are typical radiologic features of retroperitoneal leiomyosarcoma (LMS). Classically, retroperitoneal LMS is seen as large tumors containing necrotic areas with vascular invasion on cross-sectional imaging (3). E-GIST was not considered in the differential diagnosis in the first place and a diagnostic challenge in the current case due to the venous involvement and retroperitoneal localization of the tumor.

In conclusion, E-GIST is an extremely rare tumor of the retroperitoneal region. Radiologically, it may resemble retroperitoneal LMS. Because the approaches and managements are quite different, histopathological and immunohistochemical studies are essential for differential diagnosis.

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**Informed Consent:** Written informed consent was obtained from patient who participated in this case.

**Peer-review:** Externally peer-reviewed.

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## REFERENCES

1. Al-Jehani YM, Bousbait HI, Kant BR. Extra-gastrointestinal stromal tumor presenting as a surgical emergency. Turk J Gastroenterol 2011; 22: 617-20.
2. Casella C, Villanacci V, D'Adda F, Codazzi M, Salerni B. Primary Extra-gastrointestinal Stromal Tumor of Retroperitoneum. Clin Med Insights Oncol 2012; 6: 189-7. [CrossRef]
3. Rajiah P, Sinha R, Cuevas C, Dubinsky TJ, Bush WH Jr, Kolokythas O. Imaging of uncommon retroperitoneal masses. Radiographics 2011; 31: 949-76. [CrossRef]