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Case Report

Retroperitoneal gastrointestinal stromal tumor: A case report and literature review [☆]

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ABSTRACT

Retroperitoneal gastrointestinal tumor is the rarest subtype among 3 subtypes of extragastric stromal tumors, which are uncommon stromal tumors. We herein report a case of a 55-year-old man with retroperitoneal gastrointestinal tumor detected by magnetic resonance imaging and confirmed by postoperative histology and immunohistochemistry.

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Introduction

Gastrointestinal stromal tumors (GISTs) are nonepithelial neoplasm located in the muscular layer of the gastrointestinal (GI) tract [1]. They are thought to be originated from GI pacemaker cells named interstitial cells of Cajal. The incidence is approximately 14.5 patients per 1 million persons [2,3]. Extragastric stromal tumors (EGISTs) are a group of tumors arising from outside of the digestive tract, ie,

in the omentum, mesentery, prostate, retroperitoneum... [4,5]. Although sharing similar histology and immunohistochemical features with GISTs, EGISTs show different characteristics in term of clinicopathology, radiology as well as prognosis and treatment [4]. EGISTs are very rare, accounting for less than 1% of malignant GI tumors and around 10% of all GISTs [2,6].

We report a case of a 55-year-old man with retroperitoneal EGIST confirmed by postoperative histological and immunohistochemical results.

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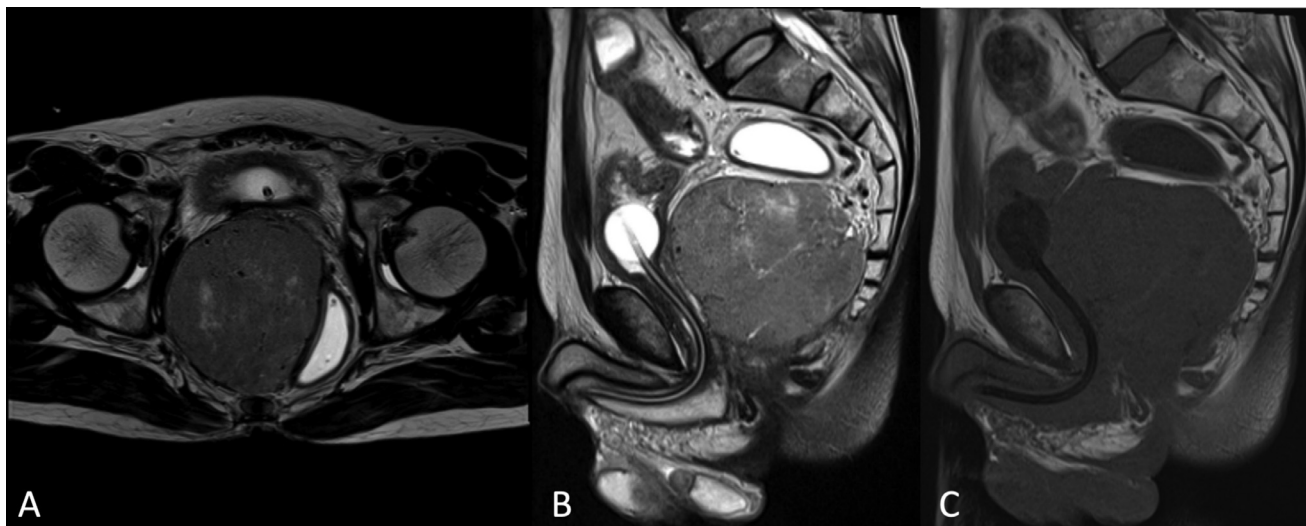


Fig. 1 – Axial T2W (A), sagittal T2W (B), and sagittal T1W (C) images show a large lobulated mass in the pelvic cavity, located between the posterior neck of urinary and the anterior rectal wall.

Case report

A 55-year-old male patient was admitted to our hospital with acute urinary retention. He had complaints of progressive dysuria and mild constipation for 2 weeks. Clinical examination did not detect any palpable abdominal mass. Digital rectal examination revealed a mass in the right anterior quadrant of the rectal wall, located 1.5 cm above the dentate line. The mass was solid, immobile with smooth surface. No blood was found on the exploratory finger. All laboratory tests were within normal limits.

Pelvic magnetic resonance imaging (MRI) revealed an 8.5 × 8 × 7.4cm, well-defined, lobulated soft tissue mass located between the posterior neck of urinary and the anterior rectal wall. This mass was isointense on T1W images, slightly heterogeneous hyperintense on T2W images compared with the adjacent muscle (Fig. 1). It showed heterogeneously enhancement with some central cystic-necrosis components (Fig. 2). The solid portions of the mass show high-signal intensity on diffusion-weighted imaging and low apparent diffusion coefficient values, consistent with restricted diffusion (Fig. 3). There was no calcification found. The mass displaced the rectum but was not separable from the distal rectal wall. It was in touch with the posterior border of the prostate, displaced prostate anteriorly, and both seminal vesicles superiorly. There was no evidence of adjacent infiltration or lymph node metastasis.

The patient underwent a complete en bloc removal of the mass. During performing surgery, the mass was easily resected from the right ureter, anterior rectal wall, and the surrounding organs. Histologically, the tumor consisted of spindle cells. Immunohistochemical result showed that the tumor cells were negative for desmin and smooth muscle actin. The tumor cells only expressed vimentin, CD117, CD34, S100. There was mildly positive for Ki67 sporadic (50%). The immunohistochemical and histologic examinations revealed

a stromal tumor with high risk (8.5 cm, 10/50 HPF), according to 2006 Miettinen and Lasota classification (Figs. 4-11) [7].

Discussion and conclusion

GISTs are the most common mesenchymal tumors of GI tract, accounting for approximately 1% of all GI malignancies [8,9]. The term EGIST was first named by Reith et al in 2000, describing all tumors that raised outside the GI tract sharing similar clinical presentation, imaging, histopathological, and molecular features with GISTs [10]. EGISTs are composed of 3 subtypes with retroperitoneal EGISTs is the rarest among them [4,8]. They are predominantly found in adults, ranging from 50 and 60 years old [11]. The clinical symptoms of GISTs and EGISTs vary according to the location and size of tumors [12,13]. In histological studies, tumor cells in GISTs are usually spindle or epithelioid-shaped. Immunohistochemically, GISTs are positive for c-KIT (CD117) and CD34 but negative for desmin. They also showed variable positivity for smooth muscle actin while positive results for S100 were found rare [7,14].

The imaging findings of stromal tumors are variable. Generally, tumors are known as well-defined masses originating from GI wall. They are often heterogeneous, with intratumoral hemorrhage, cystic change, or necrosis [9,14]. Radiological characteristics of primary GISTs include large mass (size >5 cm), exophytic growth, heterogeneous enhancement, central cyst or necrosis, mucosal ulceration, neoplastic vessels, and aneurysm. A peripheral enhancement pattern with central cystic or necrosis nonenhancement is considered indicative of GISTs. Lymphadenopathy does not appear to be a part of GISTs. Calcification, ascites and hemorrhage are also infrequent. Radiologic characteristics of EGISTs in some studies identify as a large mass, with solid and cystic components, heterogeneous enhancement [11,14]. On computed

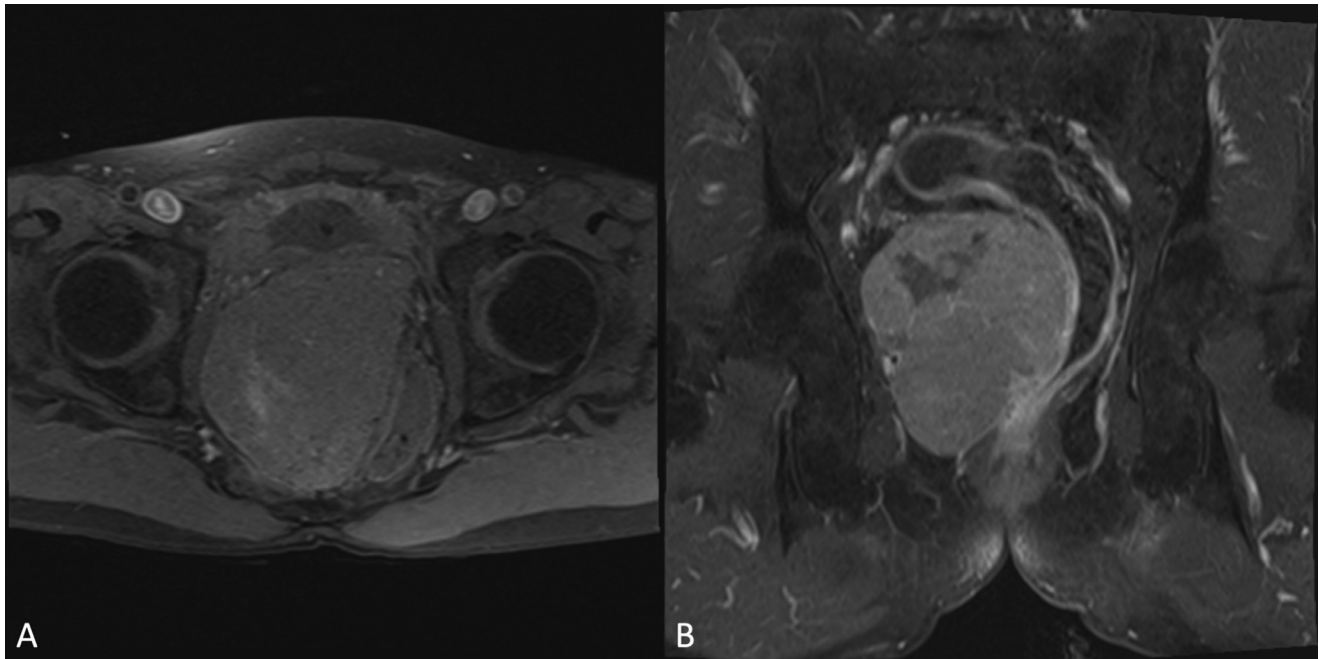


Fig. 2 – Fat-saturated axial T1W (A) and coronal T1W (B) after intravenous Gadolinium administration. The mass showed heterogeneous enhancement with nonenhanced central cystic-necrosis component.

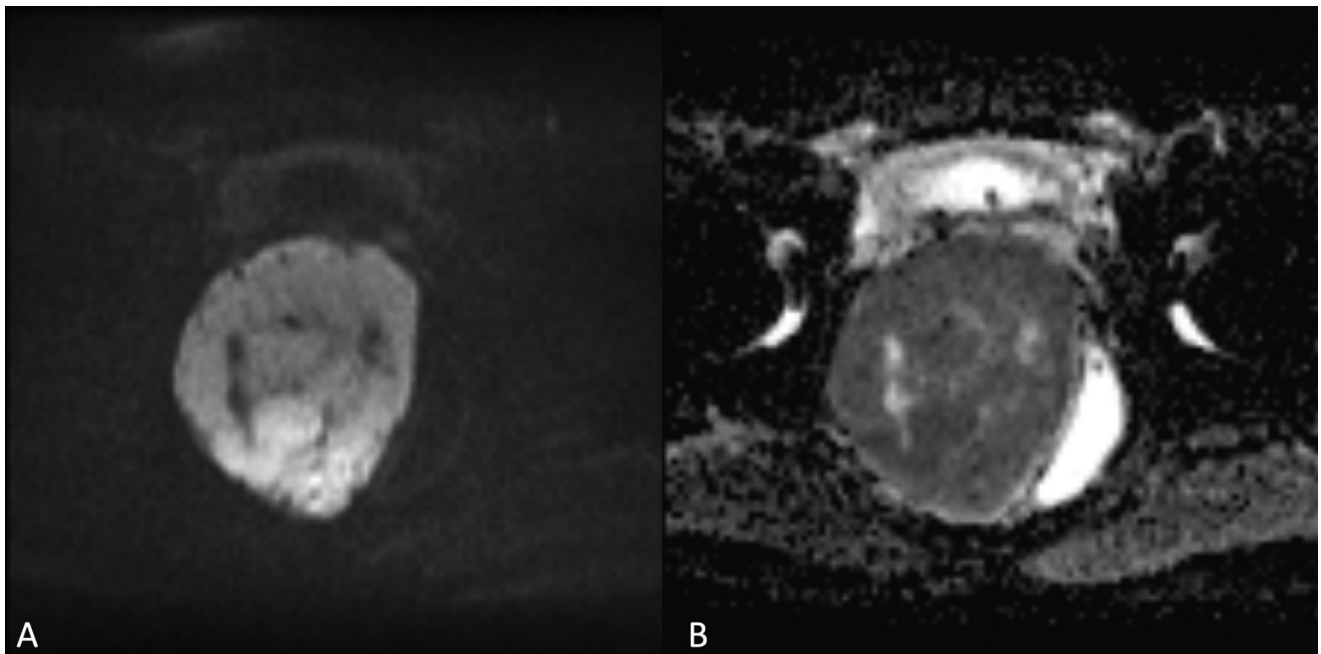


Fig. 3 – The solid portions of the mass have high-signal intensity on DWI (A) and low ADC values (B), consistent with diffusion restriction.

tomography, GISTs usually have heterogeneous soft tissue density. While cystic fluid or necrosis is familiar, calcification is rare. GISTs typically show strong enhancement [13]. On MRI, solid portions show hypointensity on T1W images, hyperintensity on T2W images, and enhance vividly after intravenous administration of gadolinium. There might be

restricted diffusion [14]. Cystic degeneration usually occurs in large masses due to the hyperplastic feature of the tumor [9].

Differential diagnosis between EGISTs and other types of tumors, for instance, lymphoma, leiomyoma, leiomyosarcoma, fibrosarcoma... is problematic because of their overlapped appearance [8,11]. Differential diagnosis also includes

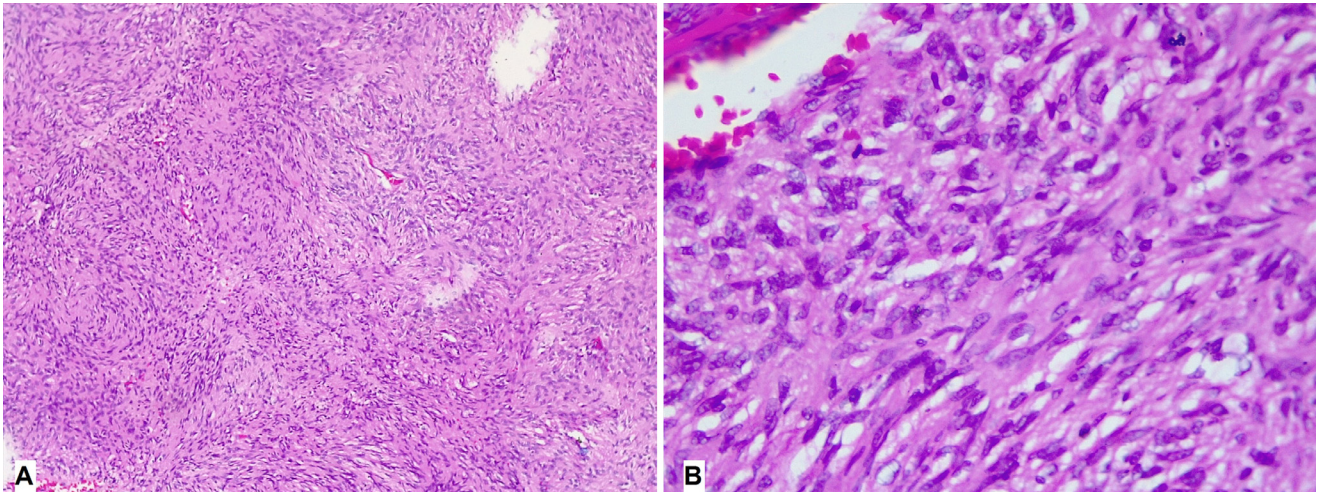


Fig. 4 – Histological result shows well-defined spindle cells with eosinophilic cytoplasm in a syncytial pattern, elongated nuclei with inconspicuous nucleoli; HE x40 (A) and HE x400 (B).

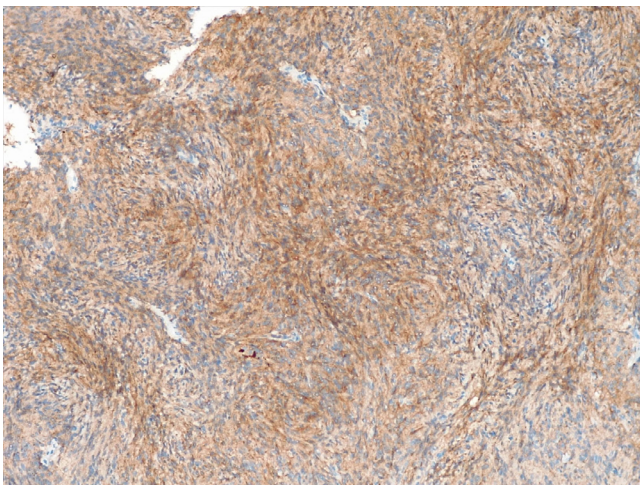


Fig. 5 – Immunohistochemical result shows positive for CD117, IHC x40.

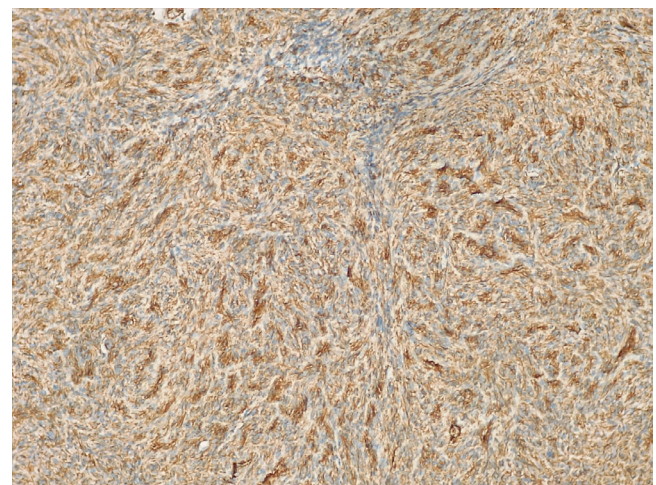


Fig. 6 – Positive for vimentin, IHC x40.

finding the primary site of GISTs. Based on the characteristics of tumor in our case, we had to clarify that retroperitoneal tumors such as prostate cancer, rectal carcinoma invaded retroperitoneum or, if the GIST arise from the rectum, prostate with other organs invasion.

To our knowledge, retroperitoneal EGISTs are considered rare, with only 60 cases reported in the English literature. Therefore, clinical findings, imaging characteristics, and management for this type of tumor have not been totally elucidated [8]. Computed tomography plays an important role in the detection and diagnosis of GISTs, evaluation of adjacent structure invasion, local and distal metastasis... MRI, with characteristics of high soft tissue contrast and multiplanar reconstruction, is becoming popular in the use of determining the primary site of large tumors and finding the relationship between the mass and major blood vessels, as well as surrounding organs [9]. However, due to their irregularity and

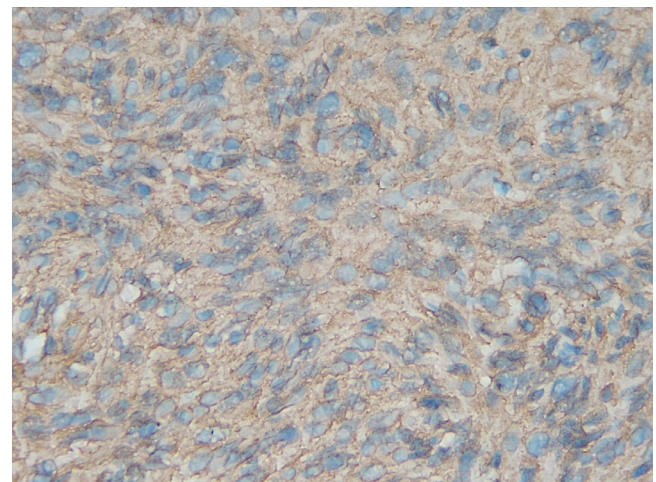


Fig. 7 – CD34 positive, IHC x400.

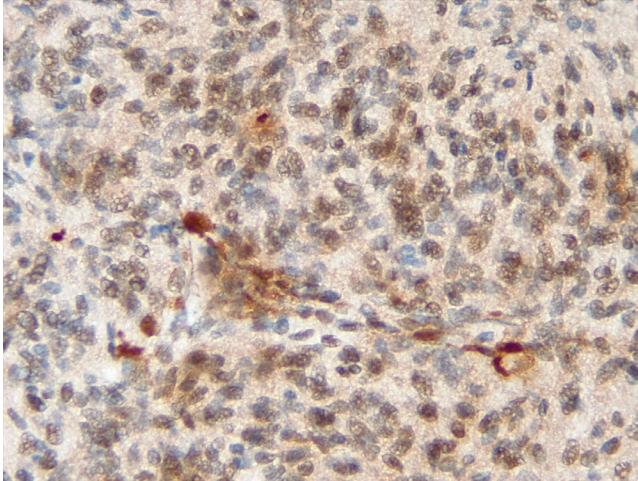


Fig. 8 – S100 positive, IHC x400.

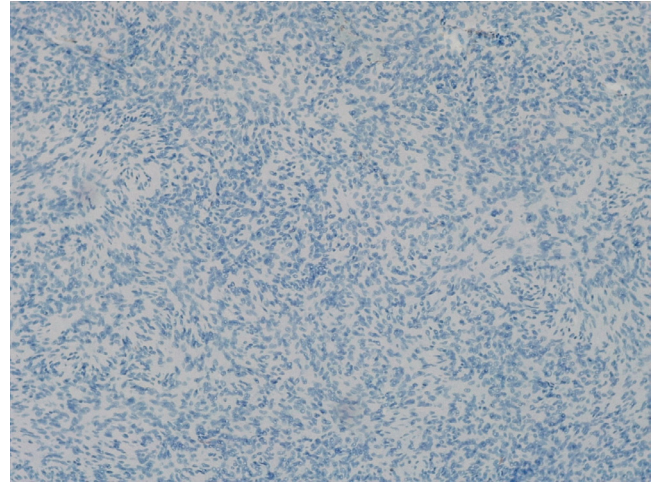


Fig. 11 – Negative for SMA, IHC x40.

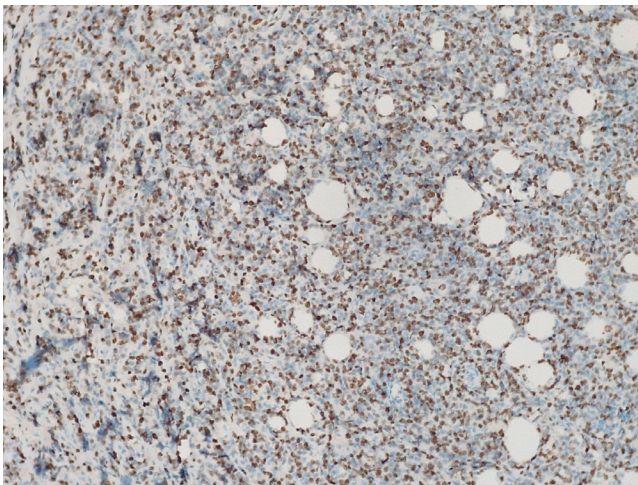


Fig. 9 – Mildly positive of Ki67 sporadic consistent with the high risk of malignancy (often >14%), IHC x100.

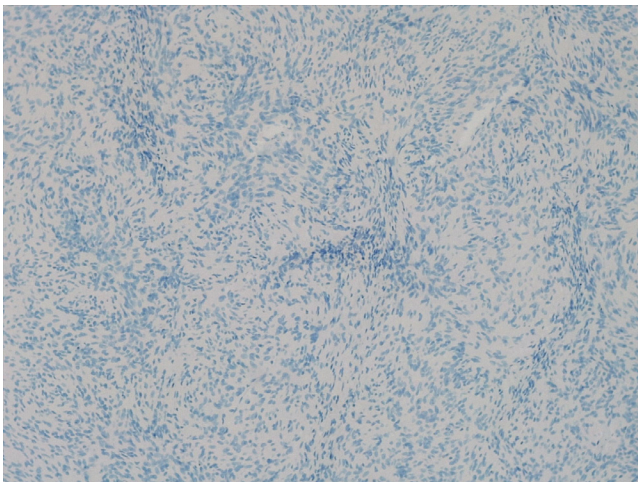


Fig. 10 – Desmin negative, IHC x40.

absence of typical features, the definitive diagnosis can only be confirmed after biopsy or surgery, with positive result from immunohistochemistry of c-KIT (CD 117), a tyrosine kinase growth factor receptor [7,8].

In conclusion, even being a rare entity, EGISTs should be included in the differential diagnosis of retroperitoneal masses, indicating the need for further studies to propose typical radiological features of this type of tumor.

REFERENCES

- [1] Park S-S, Min B-W, Kim W-B, Choi J-W, Lee J-H, Chae Y-S, et al. Malignant extragastrointestinal stromal tumor of retroperitoneum. *Acta Oncol* (Stockholm, Sweden) 2005;44:497–9. doi:10.1080/02841860510029897.
- [2] Cho M-Y, Sohn JH, Kim JM, Kim K-M, Park YS, Kim WH, et al. Current trends in the epidemiological and pathological characteristics of gastrointestinal stromal tumors in Korea, 2003-2004. *J Korean Med Sci* 2010;25(6):853–62. doi:10.3346/jkms.2010.25.6.853.
- [3] Sasmal PK, Sharma R, Patra S, Mishra TS, Mishra P, Rout B. Malignant extra-gastrointestinal stromal tumor of the mesentery. *Surg J* (N Y) 2019;5(3):e65–8. doi:10.1055/s-0039-1693040.
- [4] Laroia ST, Yadav T, Rastogi A, Sarin S. Malignant retroperitoneal extra-gastrointestinal stromal tumor: a unique entity. *World J Oncol* 2016;7(2–3):45–50. doi:10.14740/wjon926w.
- [5] Nguyen Thanh T, Nguyen TTN, Le TB, Le DD, Nguyen VM, Le DK. Extragastrointestinal stromal tumor presenting as an exophytic prostatic mass. *Radiol Case Rep.* 2020;15(8):1142–8. doi:10.1016/j.radcr.2020.05.003.
- [6] Yi JH, Park B-B, Kang JH, Hwang IG, Shin DB, Sym SJ, et al. Retrospective analysis of extra-gastrointestinal stromal tumors. *World J Gastroenterol* 2015;21(6):1845–50. doi:10.3748/wjg.v21.i6.1845.
- [7] Miettinen M, Lasota J. Gastrointestinal stromal tumors: pathology and prognosis at different sites. *Semin Diagn Pathol* 2006;23(2):70–83. doi:10.1053/j.semdp.2006.09.001.
- [8] Costa Almeida C, Caroco TV, Albano M, Carvalho L. Extragastrointestinal stromal tumour (EGIST) presented as a

- mesenteric and retroperitoneal mass. *BMJ Case Rep* 2019;12(12). doi:[10.1136/bcr-2019-232481](https://doi.org/10.1136/bcr-2019-232481).
- [9] Yu MH, Lee JM, Baek JH, Han JK, Choi B-I. MRI features of gastrointestinal stromal tumors. *Am J Roentgenol* 2014;203(5):980–91. doi:[10.2214/AJR.13.11667](https://doi.org/10.2214/AJR.13.11667).
- [10] Reith JD, Goldblum JR, Lyles RH, Weiss SW. Extragastrintestinal (soft tissue) stromal tumors: an analysis of 48 cases with emphasis on histologic predictors of outcome. *Mod Pathol* 2000;13(5):577–85. doi:[10.1038/modpathol.3880099](https://doi.org/10.1038/modpathol.3880099).
- [11] Zhu J, Yang Z, Tang G, Wang Z. Extragastrintestinal stromal tumors: computed tomography and magnetic resonance imaging findings. *Oncol Lett* 2015;9(1):201–8. doi:[10.3892/ol.2014.2705](https://doi.org/10.3892/ol.2014.2705).
- [12] Chen Y-C, Tsai J-W, Chen H-P, Lin I-C, Lin VC. Retroperitoneal extra-gastrointestinal stromal tumor presenting with lower urinary tract symptoms. *Urol Sci* 2010;21(3):148–51. doi:[10.1016/S1879-5226\(10\)60033-4](https://doi.org/10.1016/S1879-5226(10)60033-4).
- [13] Michael King D. The radiology of gastrointestinal stromal tumours (GIST). *Cancer Imaging* 2005;5(1):150–6. doi:[10.1102/1470-7330.2005.0109](https://doi.org/10.1102/1470-7330.2005.0109).
- [14] Levy AD, Remotti HE, Thompson WM, Sobin LH, Miettinen M. From the archives of the AFIP. *RadioGraphics* 2003;23(2):283–304. doi:[10.1148/rg.232025146](https://doi.org/10.1148/rg.232025146).