Radiology Case Reports 16 (2021) 2315–2318

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case report

A case of KIT negative extra-gastrointestinal stromal tumor arising in the greater omentum with predominant cystic formation ☆,☆☆

Katsuya Nanjo, MD[a], *Yusuke Nishimuta, MD*[b], *Mitsutoshi Miyasaka, MD*[b], *Kenji Shinozaki, MD, PhD*[a], *Daisuke Tsurumaru, MD, PhD*[c,*], *Kousei Ishigami, MD, PhD*[c]

[a] *Department of Radiology, National Hospital Organization Kyushu Cancer Center, 3-1-1 Notame, Minami-ku, Fukuoka, 811-1395, Japan*

[b] *Department of Gastrointestinal Endoscopy, National Hospital Organization Kyushu Cancer Center, 3-1-1 Notame, Minami-ku, Fukuoka, 811-1395, Japan*

[c] *Department of Clinical Radiology, Graduate School of Medical Sciences, Kyushu University, 3-1-1 Maidashi, Higashi-ku, Fukuoka, 812-8582, Japan*

ARTICLE INFO

Article history:
Received 22 May 2021
Accepted 23 May 2021

Key Words:
Gastrointestinal stromal tumor
Extra-gastrointestinal stromal tumor
KIT
Greater omentum

ABSTRACT

We report a rare case of KIT-negative extra-gastrointestinal stromal tumor, in a 40-year-old woman. Contrast-enhanced computed tomography and magnetic resonance imaging revealed a >15-cm mass of multiple cystic lesions in the greater omentum. Histopathological findings after surgery showed a sheet-like growth of stellate tumor cells from epithelial cells, cystic degeneration, and mucus-like stroma. Immunohistochemistry was positive for discovered on GIST-1 (DOG1) but negative for CD117 (c-kit).

Introduction

Gastrointestinal stromal tumor (GIST) is the most common mesenchymal neoplasm of the gastrointestinal tract and is this type of GIST generally has low mitotic activity and a relatively favorable prognosis [3,4].

A tumor that manifests GIST-like features located at extra-gastrointestinal regions was described as an extra-gastrointestinal stromal tumor (EGIST), and it was reported

☆ Acknowledgments: The authors acknowledge Drs. Sawamoto Hirofumi, Jinnouchi Mikako, Kusumoto Chie, Watanabe Hirofumi, Department of Radiology, National Hospital Organization Kyushu Cancer Center, for analyzing image findings for this manuscript. The authors acknowledge Dr. Taguchi Kenichi, Department of Pathology, National Hospital Organization Kyushu Cancer Center, for providing the pathological information and Ohta Mitsuhiko, Department of Gastroenterological Surgery, National Hospital Organization Kyushu Cancer Center, for providing the clinical information.

☆☆ Competing Interest: None

* Corresponding author: D.T
E-mail address: tsurumaru.daisuke.931@m.kyushu-u.ac.jp (D. Tsurumaru).

https://doi.org/10.1016/j.radcr.2021.05.061

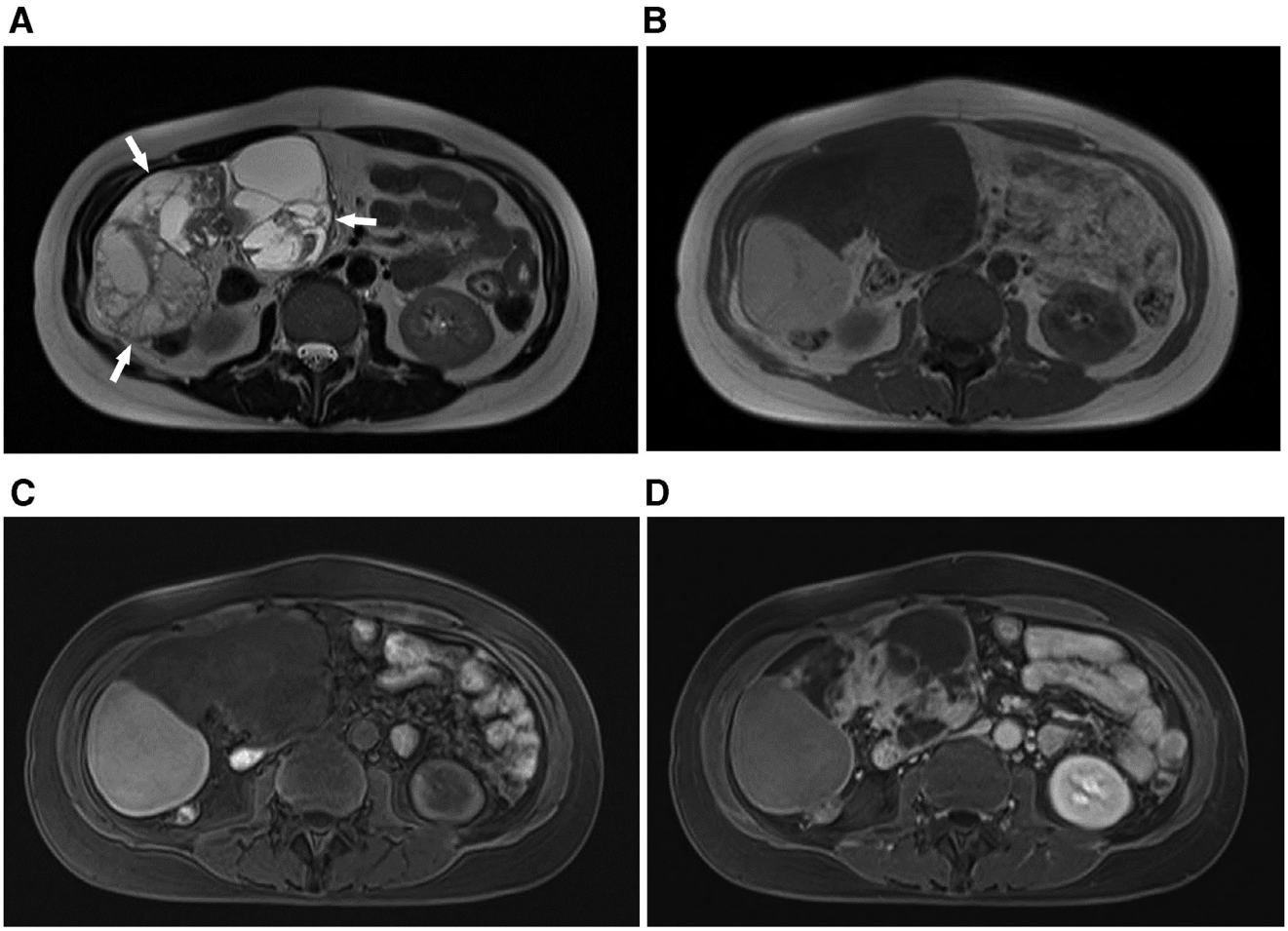

Fig. 1 – (A) MRI clearly revealing the patient's multilocular cystic mass with very high intensity on the T2-weighted image. The signal in the cysts was variable, and some of the cysts showed high intensity on T1-weighted imaging (B) with fat suppression (C), suggesting blood or viscous protein components. (D) Gadolinium-enhanced T1-weighted image with fat suppression showing heterogeneous contrast enhancement at the irregularly thickened walls and septum.

that EGISTs most often occur in the omentum, mesentery, or retroperitoneal space [5]. KIT-negative EGISTs are extremely rare, and the clinicopathological and radiological features have not been well documented [4,6,7]. Here, we report a case of KIT-negative EGIST that arose in the greater omentum, forming a large mass with predominantly cystic changes.

Case report

A 40-year-old woman with the complaint of an abdominal mass visited our institution. Laboratory tests indicated anemia with a hemoglobin value of 8.9 g/dL. The levels of tumor markers including carcinoembryonic antigen, carbohydrate antigen 19-9, cancer antigen 125, α-Fetoprotein, and squamous cell carcinoma antigen were within the normal ranges.

Contrast-enhanced abdominal computed tomography (CT) revealed a large mass (15 cm in dia.) consisting of multiple cystic lesions located in the greater omentum. The tumor showed heterogeneous contrast enhancement at the irregularly thickened walls and septum. There were no signs of liver or peritoneal metastases. Magnetic resonance imaging (MRI) clearly showed a multilocular cystic mass with very high intensity on the T2-weighted image (Fig. 1A) The signal in the cysts was variable, and some of the cysts showed high intensity on T1-weighted images (Fig. 1B,C), suggesting blood or viscous protein components. A gadolinium-enhanced T1-weighted image with fat suppression also showed heterogeneous contrast enhancement at the irregularly thickened walls and septum (Fig. 1D). Single-shot echo-planar diffusion-weighted imaging (DWI) ($b = 800$ s/mm^2) and an apparent diffusion coefficient map showed the solid part of the tumor with visually assessed diffusion restriction. These radiological findings indicated a multicystic tumor of the greater omentum with malignant potential, such as cystic mesothelioma, mucinous cystic tumor, peritoneal carcinoma.

We performed a tumor resection. The surgical findings showed a multifocal cystic mass in the greater omentum with no invasion to surrounding organs, and the mass was successfully removed. Microscopically, the tumor showed a proliferation of epithelioid to stellate tumor cells with round to oval-

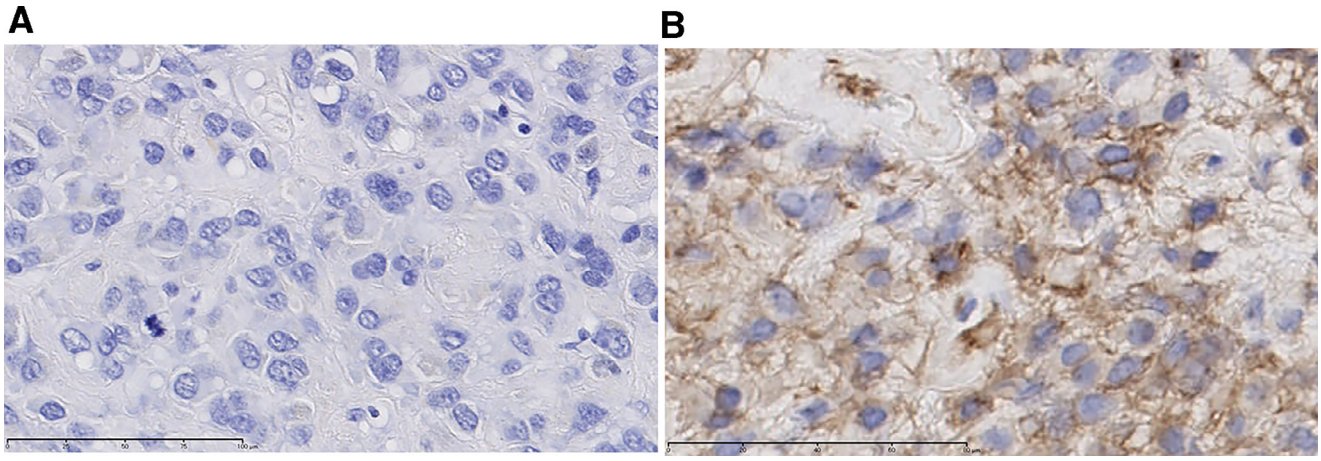

Fig. 2 – Microscopically, the tumor showed a proliferation of epithelioid to stellate tumor cells with round to oval-shaped vesicular nuclei. The tumor cells were immunohistochemically positive for discovered on GIST-1 (DOG1) (A) but negative for c-kit (CD117) (B).

shaped vesicular nuclei and clear to eosinophilic cytoplasm with intracytoplasmic vacuoles arranged in sheet-like patterns, accompanied by cystic formations and myxoid stroma. Only a few mitotic figures including atypical mitosis were observed.

The tumor cells were immunohistochemically positive for p16, discovered on GIST-1 (DOG1) (Fig. 2A), MDM2, D2-40, desmin, and H3K27me3, but negative for CAM5.2, AE1/AE3, BerEP4, smooth muscle actin (SMA), S-100, WT1, calretinin, c-kit (Fig. 2B), and brachyury. PDGFRA gene mutation (exon18) was detected by direct sequencing. We finally diagnosed the tumor as a KIT-negative EGIST originating from the greater omentum.

A tumor >10 cm with any mitotic rate is considered to be a high-risk factor based on the modified U.S. National Institutes of Health classification. However, the patient refused any adjuvant chemotherapy. She has remained alive with no recurrence for >6 months at the latest observation.

Discussion

The preoperative diagnosis of EGIST is difficult because of its non-specific clinical features, even though the tumor becomes very large and causes anemia. Several studies have reported imaging features of primary GIST [8-11]. A typical GIST shows masses arising from the gastrointestinal wall and projecting into the abdominal cavity. The imaging features vary depending on the tumor's size and aggressiveness [9]. Large tumors (>6 cm) frequently show a peripheral enhancement with central necrosis, cystic changes, or intratumoral hemorrhage [9, 11]. Zhu et al. reported CT and MRI features of EGISTs. According to their results, EGISTs showed a round or oval shape (66.7%), cystic-solid (87.5%) and ill-defined (66.7%) contour, and hypodense (69.6%) or isodense (30.4%) masses on CT; hypointensity (50%), isointensity (33.3%) or hyperintensity (16.7%) on T1-weighted images; and hyperintensity on T2-weighted images (100%) and DWI (100%) on MRI [12].

A few studies have described the imaging features of KIT-negative GISTs or EGISTs. Tateishi et al. reported that KIT-weak or KIT-negative GISTs showed large extraluminal masses with a heterogeneous lesion containing cystic and soft tissue elements on CT [7]. However, these non-specific image features overlap those of other benign or malignant intra-abdominal tumors. Advanced patient age, large tumor size, cystic-necrotic components, rare lymphadenopathy, a pattern of heterogeneous enhancement, and hepatic metastasis may aid in the diagnosis of EGIST [12]. In addition, large tumors forming multiple cystic components on CT or MRI as observed in our patient's case may be a specific imaging feature.

Authorship

(1) The conception and design of the study: Nanjo K, Nishimuta Y,
(2) Drafting the article: Miyasaka M, Shinozaki K
(3) Final approval of the version to be submitted: Tsurumaru D, Ishigami K

Patient consent statement

A formal consent is not required for the use of entirely anonymized images from which the individual cannot be identified- for example, CT, MRI images or pathology slides, provided that these do not contain any identifying marks and are not accompanied by text that might identify the individual concerned

REFERENCES

[1] Rubin BP, Heinrich MC, Corless CL. Gastrointestinal stromal tumour. Lancet 2007;369(9574):1731–41.

[2] Nishida T. Asian consensus guidelines for gastrointestinal stromal tumor: what is the same and what is different from global guidelines. Transl Gastroenterol Hepatol 2018;3:11.
[3] Medeiros F, Corless CL, Duensing A, Hornick JL, Oliveira AM, Heinrich MC, et al. KIT-negative gastrointestinal stromal tumors: proof of concept and therapeutic implications. Am J Surg Pathol 2004;28(7):889–94.
[4] Yamamoto H, Kojima A, Nagata S, Tomita Y, Takahashi S, Oda Y. KIT-negative gastrointestinal stromal tumor of the abdominal soft tissue: a clinicopathologic and genetic study of 10 cases. Am J Surg Pathol 2011;35(9):1287–95.
[5] Fagkrezos D, Touloumis Z, Giannila M, Penlidis C, Papaparaskeva K, Triantopoulou C. Extra-gastrointestinal stromal tumor of the omentum: a rare case report and review of the literature. Rare Tumors 2012;4(3):e44.
[6] Ogawa H, Gotoh K, Yamada T, Takahashi H, Ohigashi H, Nagata S, et al. A Case of KIT-Negative Extra-Gastrointestinal Stromal Tumor of the Lesser Omentum. Case Rep Gastroenterol 2012;6(2):375–80.
[7] Tateishi U, Miyake M, Maeda T, Arai Y, Seki K, Hasegawa T. CT and MRI findings in KIT-weak or KIT-negative atypical gastrointestinal stromal tumors. Eur Radiol 2006;16(7):1537–43.
[8] Levy AD, Remotti HE, Thompson WM, Sobin LH, Miettinen M. Gastrointestinal stromal tumors: radiologic features with pathologic correlation. Radiographics 2003;23(2):283–304 456quiz 532.
[9] Ghanem N, Altehoefer C, Furtwangler A, Winterer J, Schafer O, Springer O, et al. Computed tomography in gastrointestinal stromal tumors. Eur Radiol 2003;13(7):1669–78.
[10] Hong X, Choi H, Loyer EM, Benjamin RS, Trent JC, Charnsangavej C. Gastrointestinal stromal tumor: role of CT in diagnosis and in response evaluation and surveillance after treatment with imatinib. Radiographics 2006;26(2):481–95.
[11] Chourmouzi D, Sinakos E, Papalavrentios L, Akriviadis E, Drevelegas A. Gastrointestinal stromal tumors: a pictorial review. J Gastrointestin Liver Dis 2009;18(3):379–83.
[12] Zhu J, Yang Z, Tang G, Wang Z. Extragastrointestinal stromal tumors: computed tomography and magnetic resonance imaging findings. Oncol Lett 2015;9(1):201–8.