Case report

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

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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).

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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

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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 imaging (B) with fat suppression (C), suggesting blood or viscous protein components. A gadolinium-enhanced T1-weighted image with fat suppression showing heterogeneous contrast enhancement at the irregularly thickened walls and septum (Fig. 1D).

Single-shot echo-planar diffusion-weighted imaging (DWI) (b = 800 s/mm²) and an apparent diffusion coefficient map showed the solid part of the tumor with visually assessed diffusion restriction. These radiological findings indicated a multicycstic 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-shaped nuclei and abundant cytoplasm, consistent with malignant mesothelioma.
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).

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.

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].

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

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