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Case Report

Gastric Perivascular Epithelioid Cell Tumor: A Case Report

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Abstract

Background: The World Health Organization has recently classified Perivascular Epithelioid Cell Tumors (PEComa) as distinctive cells partly associated with the blood vessel walls that usually express melanocytic and smooth muscle markers. Between 2010 and 2017, six cases of gastric PEComa have been reported. Case presentation: We report the case of a 55-year-old Japanese woman with gastric PEComa in the lesser curvature. Preoperatively, the patient was clinically diagnosed with gastrointestinal stromal tumor. She had undergone a laparoscopic partial gastrectomy via endoscopic submucosal resection. Pathologically, a part of the tumor showed positive histochemistry for Melan A, alpha Smooth Muscle Actin (SMA), and desmin. Conclusions: Gastric PEComa is a very rare neoplasm; however, PEComa should be considered in the differential diagnosis of gastric neoplasia because four of seven cases of gastric PEComa have malignant potential.

Keywords: Gastrointestinal Stromal Tumor; Perivascular Epithelioid Cell Tumors; Stomach

List of Abbreviations

PEComa: Perivascular Epithelioid Cell Tumors; GIST: Gastrointestinal Stromal Tumor; CT: Computed Tomography; MRI: Magnetic Resonance Imaging; EUS-FNA: Endoscopic Ultrasound-Guided Fine Needle Aspiration; EMA: Epithelial Membrane Antigen; SMA: Smooth Muscle Actin; TFE3: Transcriptional Factor E3; GI: Gastrointestinal

Introduction

Zamboni et al. used the term Perivascular Epithelioid Cell Tumors (PEComa) to describe tumors containing perivascular epithelioid cells that are immunoreactive to melanocytic markers when a clear cell "Sugar" tumor of the lung and a PEComa of the pancreas were found in 1996 [1]. In 2002, the World Health Organization classified PEComa as distinctive cells partly associated with the blood vessel walls that usually express melanocytic and smooth muscle markers [2]. PEComa is predominantly found in the kidneys, lungs, uterus, falciform ligament, ligamentum teres, or Gastrointestinal (GI) tract [3-5]. In GI tract, PEComa is particularly found in the small intestine and colon [5-7]. Only six cases of PEComa in the stomach have been reported in the English literature [8-12]. Here, we present a case of gastric PEComa encountered in our hospital.

Case Presentation

A 55-year-old Japanese woman was admitted to the Fukuoka Sanno Hospital without any complaints. Anorexia and vomiting were not observed. The carcinoembryonic antigen and carbohydrate antigen 19-9 levels were below the normal limits. Upper GI endoscopy revealed tumor pressure at the angle of the stomach (Figure 1). Ultrasonography revealed a 3.0 x 3.0 cm tumor lesion in the upper abdomen. Noncontrast Computed Tomography (CT) showed a low-density tumor near the angle of the stomach (Figure 2). The patient did not undergo contrast-enhanced CT because of she was allergic to contrast mediums. Magnetic Resonance Imaging (MRI) showed hypointensity in T1-weighted images and hypointensity, including a part of hyperintensity, in T2-weighted images (Figure 3). We were unable to make a diagnosis although Endoscopic Ultrasound-Guided Fine Needle Aspiration (EUS-FNA) was attempted preoperatively. The patient was suspected to have GIST. On laparoscopy, the tumor was found to involve the stomach; therefore, laparoscopic partial gastrectomy was

performed via endoscopic submucosal resection 1 month after she was admitted.

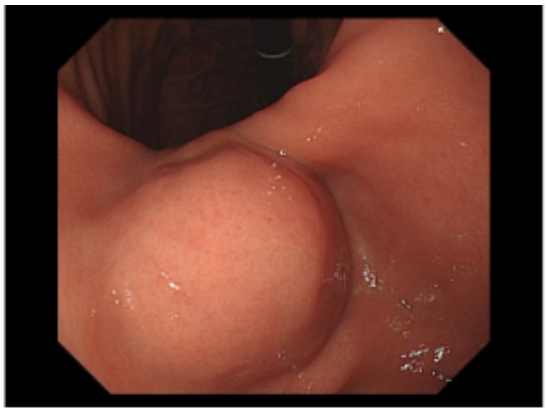

Figure 1: Gastrointestinal endoscopy showing the tumor at the angle of stomach.

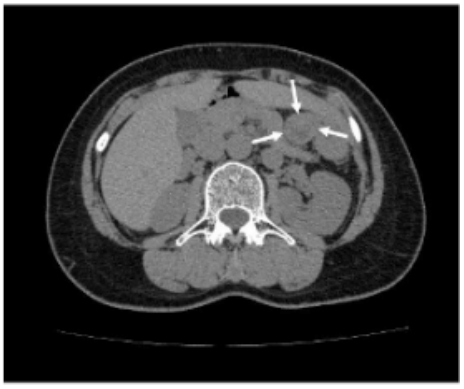

Figure 2: Computed tomography scan revealing tumors surrounding the stomach (arrow).

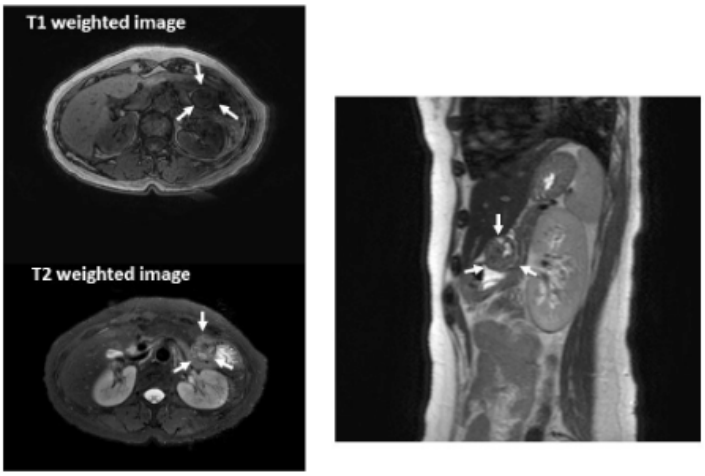

Figure 3: Magnetic resonance imaging revealed hypointensity in T1-weighted images and hypointensity, including a part of hyperintensity, in T2-weighted images near the angle of the stomach (arrow).

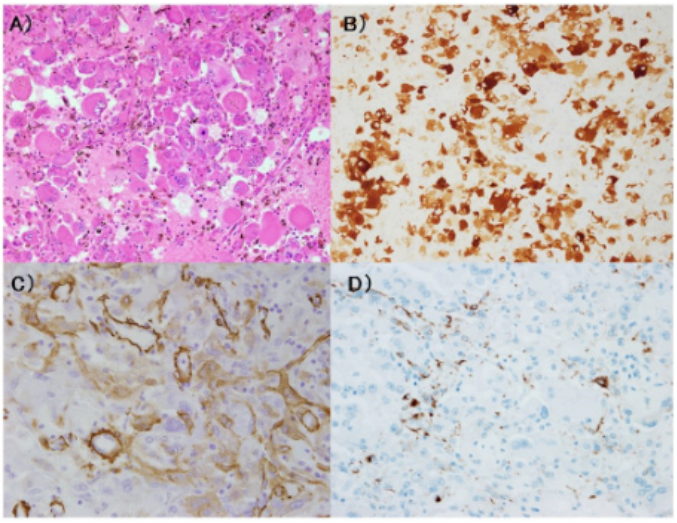

Figure 4: (A) HE stains showed a specimen of tumor. **(B)** Melan A, **(C)** alpha SMA, **(D)** desmin were positively found in the tumor specimen.

On macroscopic examination, a 3.0 x 3.0 cm mass was detected at the angle of the stomach. Therefore, a curative operation was performed. The patient was discharged on 9th postoperative day. Currently, she is doing well without recurrence at 1 year postoperatively. The patient was pathologically diagnosed with a 33 x 27 x 27 mm PEComa in the stomach characterized by a proliferation of mono- to multi-nucleated epithelioid cells with distinct small nucleoli and eosinophilic granular cytoplasm arranged in a nested or trabecular pattern from the submucosa or subserosa, accompanied by delicate capillaries, inflammatory infiltrate, hemorrhage, and cholesterol clefts (Figure 4A). Focally, spindle-shaped tumor cells were also found around the vessels. Mitotic figures are rarely observed (1/10 HPF). Immnohistochemically, the tumor cells were focally positive for Melan A, S-100 protein, EMA, alpha SMA, desmin (Figure 4B, C, D), and TFE3, but negative for HMB45, c-kit, DOG-1, AE1/AE3, synaptophysin, chromogranin A, and CD68.

Discussion

PEComa rarely involves the GI tract, and the most common sites within the GI tract are the colon and small intestine [5-7]. Only seven cases of gastric PEComa including this case have been reported in the English literature (Table 1) [8-12]. Preoperative images of PEComa do not show any specific signs. Most GI PEComa show homogenous density in plain CT and heterogenous or homogenous enhancement and, commonly, hypervascularity in contrast-enhanced CT. Usually, PEComa lesions are hypointensity to isointensity on T1-weighted MRI and heterogeneously hyperintensity on T2-weighted MRI [13]. However, abdominal CT and MRI are not sufficiently sensitive to facilitate the diagnosis of PEComa because of their nonspecific imaging characteristics.

Therefore, it is difficult to preoperatively diagnose GI PEComa. PEComa is characterized by perivascular location, and the tumor cells have mostly epithelioid and spindle appearance, with clear to lightly granular eosinophilic cytoplasm. PEC is positive for melanocytic and muscle markers [2]. Pea et al. firstly reported that renal angiomyolipomas demonstrated the presence of premelanosomes and frequent expression of HMB-45 [14].

				Microscopic feature						Immunohistochemistry			
Case	Age (yr)	Sex	Size (cm)	Infiltrative border	Nuclear grade	Mitotic activity	Necrosis	Cellularity	Prognosis	HMA-45	Melan-A	SMA	Desmin
1	71	F	3.0	-	Moderate	Low	-	NR	NED for 19 mon	-	+	+	+
2	42	M	10.0	NR	NR	NR	NR	NR	Metastasis to liver	NR	+	NR	+
3	39	M	7.3	+	High	High	+	NR	NED for 6 mon	+	+	+	+
4	48	F	11.5	NR	High	High	+	NR	-	-	+	+	NR
5	62	F	4.2	-	Low	Low	-	Low	NED for 8 mon	+	-	+	+
6	67	M	5.0	-	High	High	-	High	NED for 7 yr	-	+	+	-
Our case	55	F	3.0	+	Low	Low	+(focal)	High	NED for 1 yr	-	+	+	+

HMB-45: human melanoma black 45, SMA: smooth muscle actin, HPF: high power fields, NR: not reached, NED: no evidence of disease

Table 1: Seven cases of gastric PEComa.

On immunohistochemical analysis, most cases of PEComa are positive for melanocytic markers such as HMB-45, Melan A, microphthalmia transcription factor [4], and smooth muscle markers, such as SMA, desmin, and calponin [15]. The patient tested positive for SMA and desmin and focally positive for S-100 and TFE3, which are markers known to be found positive in a small proportion in PEComa [16]. In particular, Dole et al discussed the immunohistochemical features of PEComa in the GI tracts [5]. The immunohistochemical positive rate of desmin, Melan-A, HMB-45, microphthalmia transcription factor and SMA was >50% in the GI tracts. A consensus regarding the prognostic factors of PEComa has yet to be established because of its rarity. Approximately 100 cases of PEComa were reported and 55 of them had malignant potential [9]. We have encountered three PEComa of gynecological and renal origin in our hospital, excluding for this case. One of the four cases had recurrence after 8 years from primary surgery. Folpe et al. discussed that malignant behaviors, including PEComa recurrence or metastasis involving the soft tissues and gynecologic organs, are associated with large-sized tumors (>5 cm), frequent mitosis (>1/50 HPF), necrosis, infiltrative growth pattern, high cellularity, and high nuclear grade [4].

Other investigators have discussed that it is associated with large-sized tumors (>6 cm), frequent mitosis (>2/10 HPF), marked cellular atypia, diffuse pleomorphism, and necrosis and are diffusely infiltrative in patients diagnosed with GI tract PEComa [5]. Therefore, we believe that tumors of larger size, or those having frequent mitosis have malignant potential for GI tract PEComa. Among the seven previously reported cases of gastric PEComa, four showed histologically malignant potential [8-12]. In our case, the tumor size was 3.0 cm, and mitotic count was 1 in 10 HPE. Malignant characteristics were not observed in our case; therefore, the patient was followed-up without additional treatment. Optimal treatment is not well established because of smaller number of cases. Surgery is the most recommended primary treatment, and adjuvant therapy is generally reserved for high-risks cases. Benson et al. have retrospectively reported good response of PEComa to mTOR inhibitors (sirolimus and temsirolimus) [17]. We can consider mTOR inhibitors as adjuvant therapy in high-risks cases and those with metastatic diseases.

Conclusions

Gastric PEComa is a very rare neoplasm; however, physicians and pathologists should consider PEComa in the differential diagnosis of gastric neoplasia because four of seven cases of gastric PEComa have malignant potential. If we preoperatively suspect the diagnosis of PEComa, surgical resection is the best treatment option as obtaining a pathological diagnosis is difficult.

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Authors' Contributions

MY wrote the manuscript. MY, YY, and TG performed the surgery. MY, YY, and YT collected the data. MT, YK, and YO diagnosed the histopathological findings. All authors have read and approved the final manuscript.

Ethics Approval and Consent to Participate

Not applicable.

Consent for Publication

The informed consent for publication and presentation was obtained from the patient.

Availability of Data and Material

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

Competing Interests

The authors declared that they have no competing interests.

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