Clinical Case Report

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Imaging findings of gastric plexiform fibromyxoma with a cystic change

A case report and review of literature

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Abstract

Rationale: Plexiform fibromyxoma (PF) is an extremely rare mesenchymal tumor of the stomach, and its radiological findings have not been well described. Here, we analyzed the imaging features of a case of PF. To our knowledge, this is a rare reported case with a remarkable cystic change in the imaging literature.

Patient concerns: A previously healthy 50-year-old woman presented with a 1-day history of abdominal pain. Then, she underwent computed tomography (CT) and magnetic resonance imaging (MRI). A cystic-solid well-circumscribed extraluminal mass was located in the posterior wall of the gastric upper body. The solid portion appeared as heterogeneous attenuation/intensity with progressive enhancement while the cystic region had no enhancement.

Diagnoses: The potential for malignancy could not be excluded.

Interventions: Laparoscopic partial gastric resection was performed.

Outcomes: Based on pathological findings, a diagnosis of PF was made. The patient was alive without any recurrence or metastasis of the tumor after 2 years of follow-up.

Lessons: As far as we know, a gastric PF with a remarkable cystic change has never been reported. Additionally, the tumor exhibited a progressive enhancement pattern which is a characteristic radiographic feature in our case. Our report may help increase the awareness of this rare but important new disease entity.

Abbreviations: ADC = apparent diffusion coefficient, ALK = anaplastic lymphoma kinase, CE-T1WI = contrast enhanced T1 weighed images, CT = computed tomography, DWI = diffusion-weighted imaging, EMA = epithelial membrane antigen, F = female, GIST = gastrointestinal stromal tumors, M = male, MRI = magnetic resonance imaging, NR = not reported, PAMT = plexiform angiomyxoid myofibroblastic tumor, PF = plexiform fibromyxoma, SMA = smooth muscle actin, T1WI = T1-weighted images, T2WI = T2-weighted images, WHO = World Health Organization.

Keywords: computed tomography, cystic-solid, magnetic resonance imaging, plexiform fibromyxoma, stomach

1. Introduction

Plexiform fibromyxoma (PF) of the stomach is a recently described gastric mesenchymal tumor of the digestive tract.[1] This tumor almost occurs in the gastric antrum and seems to be a benign tumor that exhibits no recurrence, metastasis, or tumor-related mortality.[2]

The interpretation of its imaging findings and differentiation from other malignant gastric submucosal tumors is of clinical importance. To date, most reports have focused on its histology and immunohistochemistry findings. Few reports showed images of this tumor from magnetic resonance imaging (MRI) and computed tomography (CT).[5,6] Therefore, here, we would like to report a new case of cystic-solid mass that was different from other PFs because of the cystic portion with preoperative endosonography and radiological features. Furthermore, we reviewed and compared the imaging characteristics of those previously reported PFs in an effort to understand this disease better.

2. Case report

A previously healthy 50-year-old woman presented with a 1-day history of abdominal pain. The physical examination showed only mild epigastric tenderness on deep palpation without rebound. She was found to have a remarkable CA125 of 60.8 U/mL (reference range, 0–35 U/mL).

An abdominal CT (Brightspeed 16, GE Healthcare, Milwaukee) demonstrated a well-circumscribed extraluminal mass measuring 7.4 cm × 5.9 cm × 9.1 cm that was located in the posterior wall of the gastric upper body. Unenhanced CT scan showed a cystic-solid hypoattenuating mass compared with the muscle and contrast-enhanced CT scan revealed the solid portion

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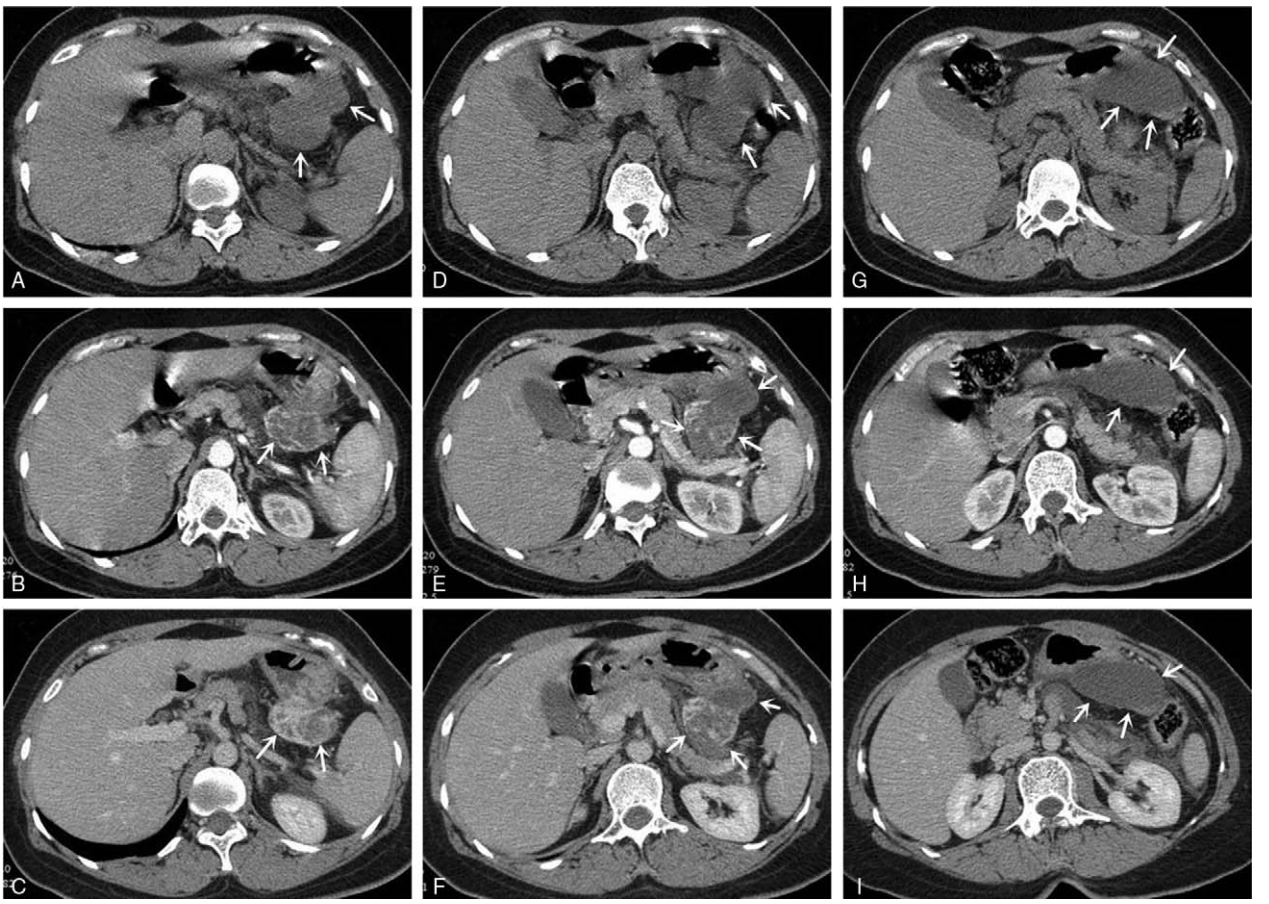

Figure 1. A 50-year-old woman with gastric plexiform fibromyxoma. A, D, G, Unenhanced CT scan showed an irregular mass (arrow) of heterogeneous hypoattenuation with the muscle that was located in the posterior wall of the gastric upper body. B, E, Contrast-enhanced CT scan revealed the solid portion (arrow) of the tumor displayed peripheral contrast enhancement in the arterial phase. C, F, The solid portion (arrow) displayed inhomogeneously gradual enhancement in the portal venous phase. H, I, The cystic region (arrow) had no enhancement. CT = computed tomography.

of the tumor displayed heterogenously gradual enhancement, while the cystic region had no enhancement (Fig. 1). The CT value of the plain scan ranged from 2.4 to 35.0 Hounsfield units (mean 18.7 Hu). Arterial phase CT showed the solid part of the tumor demonstrating peripheral mild enhancement with a CT value of 27.3 to 86.7 Hu (mean 57 Hu). In the venous phase, enhancement was strengthened progressively with a CT value of 38.7 to 128.5 Hu (mean 83.6 Hu). Arborization of the left gastric artery was observed immediately around the tumor. Moreover, a thickening of the left prerenal fascia and little fluid in the left side of the paracolic sulci were also observed. Regarding the MRI (3T Siemens Magnetom Verio, Erlangen, Germany) findings, the cystic-solid lesion was iso-intense and hypointense on the T1-weighted images (T1WI) (TR/TE, 3.25 ms/1.13 ms, slice thickness, 6 mm) and hyperintense on the T2-weighted images (T2WI) (TR/TE, 1200 ms/81 ms, slice thickness, 6 mm) (Fig. 2A1, A2, B1, B2). In particular, the cystic-solid mass was hyperintense on the diffusion-weighted imaging (DWI) (b values of 0 and 600 mm^2/s, TR/TE, 7400 ms/73 ms, slice thickness, 6 mm), in contrast to its hyperintense presentation on the apparent diffusion coefficient (ADC) map (ADC value = $3.535 \times 10–3\ mm^2/s$) (Fig. 2A3, A4, B3, B4). Contrast enhanced T1 weighed images (CE-T1WI) (TR/TE, 3.25 ms/1.13 ms, slice thickness, 4 mm) were acquired after intravenous bolus injection of gadolinium contrast agent (gadopentetate dimeglumine, Bayer Schering Pharma AG, Berlin, Germany). A lesion with heterogeneously gradual enhancement of the solid portion and a lack of obvious enhancement of the cystic region was revealed. (Fig. 2A5, A6, B5, B6). CT and MRI images did not show any evidence of gastric fistula, liquefactive necrosis, infiltration of other organs, or lymphadenopathy.

The gastroscopy showed a submucous eminent lesion in the posterior wall of the gastric upper body (Fig. 3A). The endoscopic biopsy was non-contributive. The endosonography indicated a heterogeneous hypoechoic submucosal mass that originated from the muscularis propria at the greater curvature side of the upper body. The size of the irregular lesion was approximately 5.6 cm × 3.5 cm (Fig. 3B, C).

As the potential for malignancy could not be excluded, laparoscopic partial gastric resection was performed. Gross examination of the stomach showed a large mass with envelope in the greater curvature of the upper body measuring 15 cm × 10 cm. The cystic portion was full of mucinous fluid, while the solid portion was a mucoid, gelatinous, and hemorrhagic extramural mass. The cut section showed that the tumor was mainly in the submucosa, exhibiting mucoid and hemorrhagic areas. However, no tumor nodules presented in the omental fat or distant peritoneal sites in the patient.

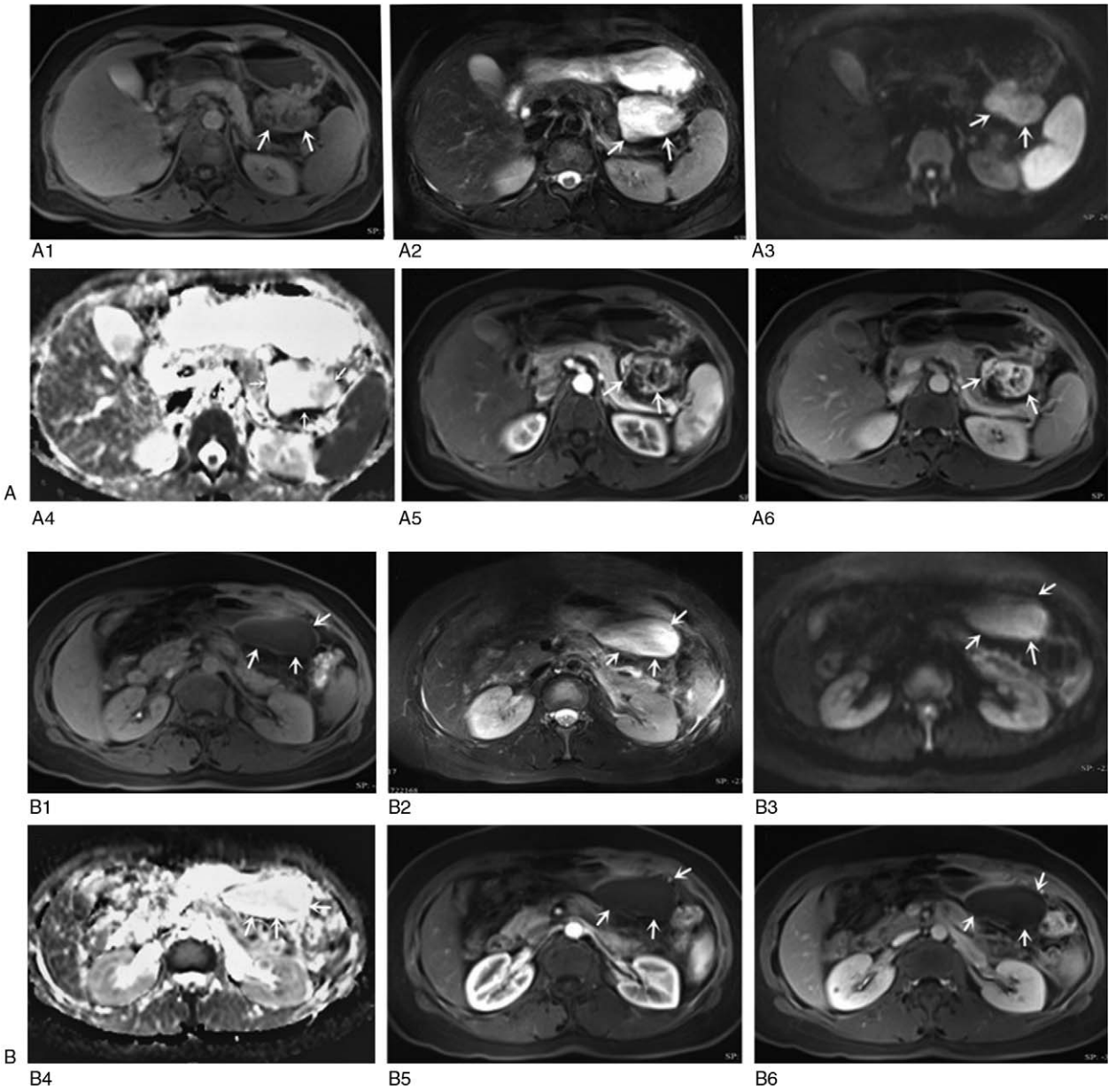

Figure 2. MRI revealed a large exophytic cystic-solid tumor (arrow) that developed from the upper body of the stomach. A1, A2 (A1, T1-weighted imaging; A2, T2-weighted imaging): the solid portion (arrow) of the tumor typically showed iso-intensity on the T1-weighted images and high intensity on the T2-weighted images. B1, B2 (B1, T1-weighted imaging; B2, T2-weighted imaging): the cystic region (arrow) displayed low and markedly high intensity on the T1-weighted and T2-weighted sequences, respectively. A3, B3 (diffusion-weighted magnetic resonance imaging): imaging techniques showed a hyperintense mass (arrow) in the stomach. A4, B4 (apparent diffusion coefficient map): the cystic-solid tumor had hyperintense presentation on ADC map. A5, A6, Contrast-enhanced T1-weighted imaging (A5 arterial phase; A6 delayed phase) demonstrated the solid portion (arrow) with heterogeneously gradual enhancement. B5, B6, Contrast-enhanced T1-weighted imaging (B5 arterial phase; B6 delayed phase) showed the cystic region (arrow) with no enhancement.

Histologic examination of the thickened wall showed tumor tissue extending from the muscularis propria up to the serosa of the stomach exhibited a multinodular plexform growth pattern. These nodules consisted of bland-looking spindle cells that were admixed with abundant myxoid or fibromyxoid stroma rich in capillary-sized vessels (Fig. 4). The component of the cystic portion was full of mucinous fluid and the wall of the cystic region was fibrous tissue. The tumor cells had no significant nuclear atypia or mitosis. No necrosis, calcification, or vascular invasion was observed. The lymph nodes showed reactive changes.

Immunohistochemically, the tumor cells were positive for smooth muscle actin (SMA), CD34, and vimentin but were negative for CD117, CD10, Dog-1, S-100 protein, desmin, cytokeratin, epithelial membrane antigen (EMA), bcl-2, HMB45, anaplastic lymphoma kinase (ALK), and β-catenin. The Ki-67 labeling index was approximately 3%.

Based on the histological features, and supported by the immunohistochemical findings, a diagnosis of PF was made. The patient was alive without any recurrence or metastasis of the tumor after 2 years of follow-up.

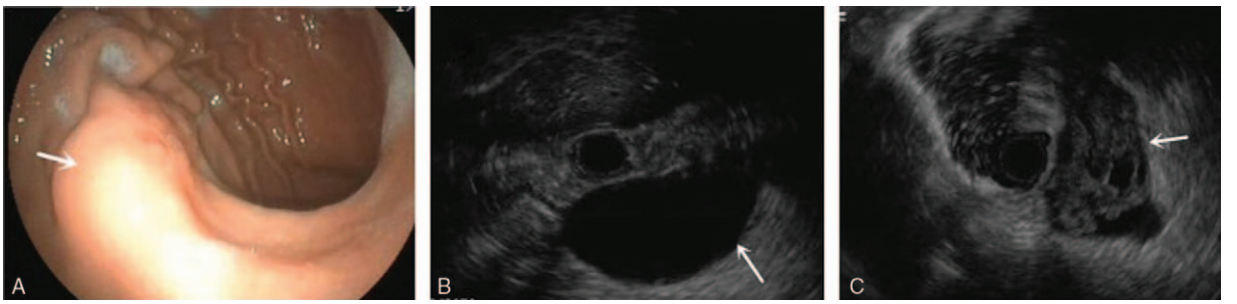

Figure 3. A, The gastroscopy showed a submucous eminent lesion (arrow) in the posterior wall of the gastric upper body. B, C, A heterogeneously hypoechoic lesion (arrow) was found at the greater curvature side of the upper body. The irregularly submucosal tumor (arrow) originated from the muscularis propria.

The ethical approval was not necessary, because it was just a case report and the study needn't involve patient consent.

3. Discussion

Plexiform fibromyxoma (PF) is a rare neoplasm of the stomach that is characterized by a peculiar plexiform growth pattern of bland spindle cells in the myxoid or fibromyxoid stroma and a delicate capillary network.[1,2,4] Since it was first reported by Takahashi et al[1] in 2007, a total of 48 cases within 57 lesions have been reported in the English-language literature under the appellation of "plexiform angiomyxoid myofibroblastic tumor (PAMT)," "plexiform angiomyxoid tumor" or "plexiform fibromyxoma (PF)."[1–22] Although the exact name is disputed, the World Health Organization (WHO) has adopted the PF designation[3] that was coined by Miettinen et al.[2] According to previous reports, these tumors were exclusively in the stomach and principally involved the submucosa and muscularis propria,[1–21] with 1 possible exception in the colon.[22] PF predominantly occurred in the gastric antrum, especially in the pyloric region.[2] Of reported cases the tumor size ranged from 1.5 to 15 cm (mean 5.7 cm). The age range of patients was 7 to 75 years (mean 41.5 years). The male-to-female ratio was approximately 1:1.[1–22] Patients often presented with nonspecific gastrointestinal symptoms, and some cases were discovered incidentally. The majority of previous series in the literature indicated that PF was biologically benign because of the bland nuclear features, low proliferative index, and absence of necrosis, recurrence, or metastasis. To date, surgical resection remains the best treatment for PF patients, and these patients have an excellent prognosis after excision.

For exogastric tumors such as those reported here, CT and MRI are the most commonly used noninvasive modalities for detection, staging, and surgical planning. However, due to the rarity of PF, limited data were available in the literature regarding the radiological features of this neoplasm.[5–12] Particularly, our case included CT and MRI imaging showing the cystic-solid tumor with heterogeneously attenuation but without ulceration or calcification. Our case was different from other PFs because of the cystic portion. Based on the images, we thought the nonenhanced cystic component may be composed of different types of fluids (e.g., serous, mucinous, proteinaceous, hemorrhagic, necrotic, or mixed). The cause of the cyst is unclear, and 1 explanation can be offered: PF is characterized by bland spindle cells in the myxoid or fibromyxoid stroma and the solid portion was a mucoid, gelatinous, and hemorrhagic mass, so the cyst formed from the mucus secreted from these cells. We speculated the wall of the cystic region was fibrous tissue because of its unenhancement. According to the pathology, the cystic part contained mucinous fluid and the wall was fibrous tissue. Therefore, the imaging features of gastric PF corresponded to the pathological composition of the lesion. The most important

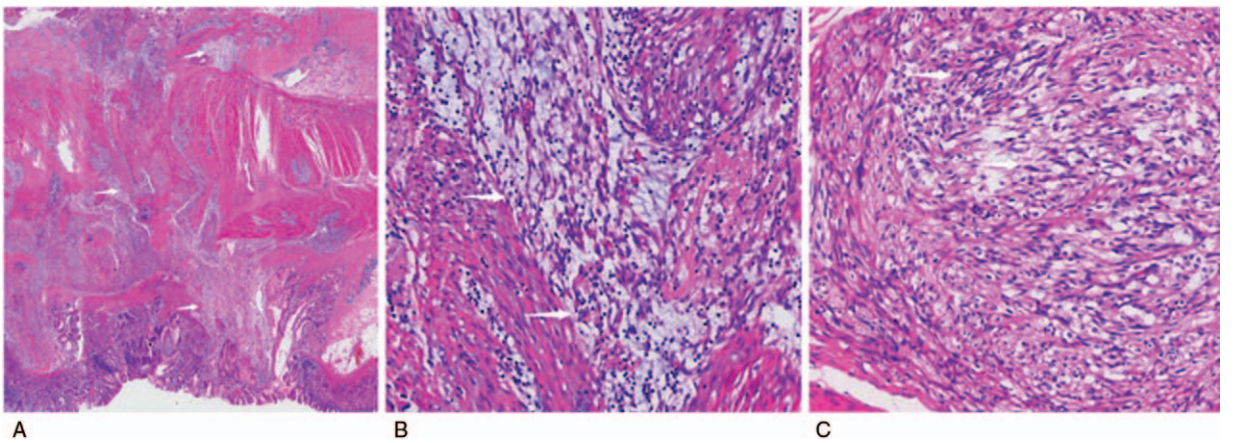

Figure 4. Pathological findings. A–C, Microscopic examination showed that the tumor extended from the muscularis propria to the serosa of the stomach and exhibited a multinodular plexiform growth pattern. These nodules consisted of bland-looking spindle cells admixed with abundant myxoid or fibromyxoid stroma rich in capillary-sized vessels.

Table 1

CT and MRI findings of the gastric PF.

Case	Sex	Age	Location	Size, cm	Component	Unenhanced CT	T1WI (MRI)	T2WI (MRI)	Enhancement pattern	Ref
1	F	50	Upper body	15	Cystic-solid	Hypo	Hypo Iso	Hyer	Heterogeneous, progressive	
2	M	60	Antrum	2	Solid	Hypo	Iso	Hyer	Heterogeneous, progressive	[5]
3	M	55	Angle	1.7	Cyst	Hypo	Iso	Hyer	Progressive	[6]
4	M	39	Antrum	3.5	Solid	Hypo	NR	NR	Heterogeneous, progressive	[7]
5	F	28	Antrum	5.5	Cystic-solid	NR	NR	NR	Heterogeneous, progressive	[8]
6	F	27	Antrum	4.6	Solid	Hypo	NR	NR	Heterogeneous, progressive	[9]
7	M	47	Midbody	3	Solid	NR	NR	NR	Prominent	[10]
8	F	55	Antrum	4	Solid	NR	NR	NR	Poor	[12]

CT = computed tomography, F = female, M = male, MRI = magnetic resonance imaging, NR = not reported.

characteristic CT feature was that the solid portion showed mild enhancement during the arterial phase and strengthened progressive enhancement during the venous and delayed phases, which was consistent with previous reports (Table 1). There was arborization of the left gastric artery around the tumor indicating a hypervascular tumor, in accordance with Lkemura.[9] According to the pathology, the gross findings of the solid portion identified a mucoid, gelatinous, and hemorrhagic mass. Therefore, the radiology findings illustrated the characteristic growth of PF, which was a hypervascular tumor located in the myxoid stroma. However, Sing et al[12] reported a case of PF with poor enhancement that we thought might be due to technological problems with the contrast-enhanced CT scan. MRI is superior to CT for visualizing tumor extent and component. In our case, gastric PF showed low signal intensity on the T1-weighted images and high signal intensity on the T2-weighted images. The solid portion also exhibited heterogeneously gradual enhancement on contrast-enhanced MR images. These MRI findings were consistent with previous reports.[5,6] The gastric PF exhibited high intensity on the diffusion-weighted imaging and the ADC map, which may be influenced by the T2 effect. In our study, pregastric lymph nodes were confirmed without evidence of neoplastic cells or malignancy by pathology.

Occasionally, some types of solid neoplasms (i.e., gastrointestinal stromal tumors, and neurogenic tumors) of the stomach appear to be cystic. Differentiating between various gastric intramural tumors may be challenging because these lesions often have overlapping radiologic appearances. The main differential diagnosis of a gastric PF is a GIST because it is the most common mesenchymal neoplasm of the stomach and a tumor >50 mm tends to have cystic degeneration, hemorrhage, and necrosis.[23] On CT and MR, the common cystic appearance of GISTs include a unilocular or multilocular thick-walled cystic-solid mass with peripheral enhancement pattern.[24] These imaging features are different from those observed in our case of gastric PF, which manifested as a well-circumscribed extraluminal cystic-solid mass with progressively heterogeneous enhancement pattern. A notable feature of schwannomas is their homogeneous attenuation. The tumor has low attenuation on unenhanced CT images and no or minimal enhancement during the arterial phase but delayed enhancement during the equilibrium phase. Although cystic degeneration is a common feature of schwannomas found in other parts of the body, this characteristic is uncommon for gastric schwannomas.[25] In our case, the mass had cystic changes which indicate that this may be helpful for differentiating gastric PF from gastric schwannomas.

To the best of our knowledge, this gastric cystic-solid PF with overall clinical and imaging findings had never been reported. In this study, the cystic-solid tumor was large with a remarkable cystic change and grew as a hypervascular exophytic mass. Additionally, the tumor exhibited a progressive enhancement pattern which is a characteristic radiographic feature in our case. Our report may help increase the awareness of this rare but important new disease entity. However, further research of a larger number of cases is needed to explore the imaging characteristics of primary PF with the goal of providing useful information for differentiating PFs from common gastric tumors in the future.

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