Clin J Gastroenterol (2015) 8:197–201
DOI 10.1007/s12328-015-0577-8

CASE REPORT

A case of a stomach gastrointestinal stromal tumor with extremely predominant cystic formation

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Received: 28 March 2015 / Accepted: 26 May 2015 / Published online: 26 June 2015

Abstract A 79-year-old male was referred to the hospital with a history of abdominal discomfort. Abdominal computed tomography revealed a cystic tumor with irregular wall thickness, approximately 50 mm in diameter, along the lesser curvature of the gastric body. Magnetic resonance imaging visualized the mass as signal-hyperintense on T2-weighted imaging. Esophagogastroduodenoscopy showed a submucosal tumor with normal mucosa. Surgery was performed and the 60 × 50 × 50 mm mass was resected. The resected tumor comprised cystic and solid regions; the cystic region was filled by light bloody serous fluid. On histological examination, a solid region of the resected tumor showed a spindle-cell appearance. The diagnosis was gastric gastrointestinal stromal tumor (GIST) with predominant cystic formation. GISTs are usually solitary tumors, but in this case the tumor demonstrated extremely predominant cystic formation. Lesions with a hemorrhage or necrosis may form large cystic spaces. GISTs may show extensive cystic changes in response to tyrosine kinase inhibitor treatment; however, this patient had not undergone any such treatment before diagnosis. This represents an interesting case of a gastric GIST with predominant cystic formation occupying most of the tumor volume. Care should be taken to differentiate between GISTs and actual intra-abdominal cystic lesions.

Keywords GIST · Cystic formation · Stomach

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Introduction

A gastrointestinal stromal tumor (GIST) is a primary non-epithelial neoplasm of the gastrointestinal tract. GISTs usually form a solid mass, sometimes presenting with focal cysts or calcification; however, GISTs rarely present with predominantly cystic changes (Table 1) [1–14]. GISTs with cystic formation are usually large and consist of a complex mass of cystic and solid lesions. Rarely, a case of a GIST with cystic degeneration and thick calcification has been reported [7]. Cystic-formed GISTs derived from the stomach, pancreas, intestine and omentum have been reported [1–14]. GISTs with predominant cystic changes may be difficult to differentiate from cystic tumors derived from other organs neighboring the gastrointestinal tract, such as pancreatic tumors. Here, we report a case of a stomach GIST with an extremely predominant relatively small cystic lesion.

Case report

A 79-year-old male was referred to our hospital with a history of abdominal discomfort. Physical and laboratory examinations yielded normal results. Abdominal computed tomography revealed a cystic tumor with irregular wall thickness, approximately 50 mm in diameter, along the lesser curvature of the body of the stomach (Fig. 1). Contrast material showed a mass with a peripheral enhancement pattern, representing enhancement of the cystic wall. Magnetic resonance imaging visualized the mass as a signal-hyperintense region on T2-weighted imaging (Fig. 2). Double-contrast gastrography showed the mass lesion protruding into the gastric lumen at the lesser curvature of the body of the stomach (Fig. 3).

Table 1 Clinical features of GISTs with cystic formation

No. of case	References	Age	Sex	Size (cm)	Origin	Mitotic index	Treatment
1	Park et al. [1]	11	Female	10	Stomach	NA	S and C
2	Osada et al. [2]	74	Male	12	Stomach	NA	S and C
3	Yang et al. [3]	55	Male	19.6	Pancreas	>150/50 HPFs	S and C
4	Cruz Jr et al. [4]	37	Male	32	Stomach	10/50 HPFs	S and C
5	Harindhanavudhi et al. [5]	63	Female	16	Pancreas	<5/50 HPFs	S and C
6	Takahashi et al. [6]	54	Male	8	Rectum	1/50 HPFs	S
7	Yu et al. [7]	81	Female	6	Stomach	4/50 HPFs	S
8	Ide et al. [8]	60 s	Male	15	Intestine	150/50 HPFs	S and C
9	Notani et al. 2013 [9]	58	Male	22	Stomach	250–500/50 HPFs	S and C
10	Shoji et al. [10]	61	Male	9	Jejunum	0/50 HPFs	S and C
11	Zhu et al. [11]	78	Male	17	Stomach	>10/50 HPFs	S
12	de AP Hansen et al. [12]	74	Female	12	Pancreas	21/50 HPFs	S
13	Ambrosio et al. [13]	72	Male	19	Pancreas	>5/50 HPFs	S and C
14		50	Female	7	Pancreas	3/50 HPFs	S
15		40	Male	7	Pancreas	15/50 HPFs	S
16	Seow-En et al. [14]	60	Female	20	Omentum	1/50 HPFs	S

S surgical resection, *C* chemotherapy, *NA* not available, *HPF* high-power field

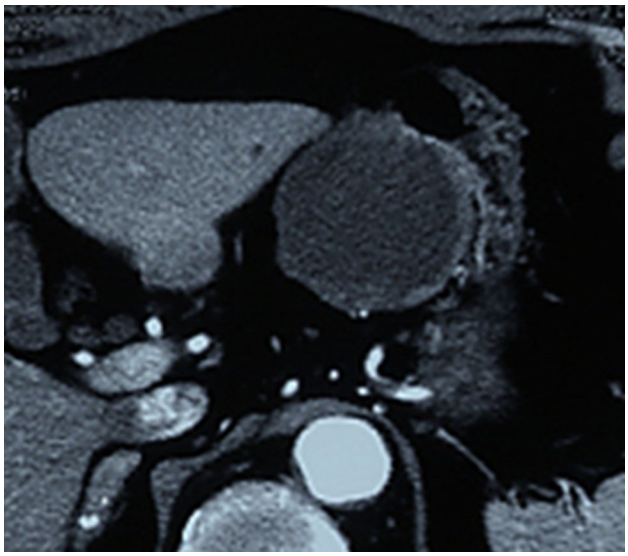

Fig. 1 Computed tomography shows a cystic tumor mass approximately 50 mm in diameter along the stomach wall. The peripheral wall of the cystic tumor shows contrast enhancement

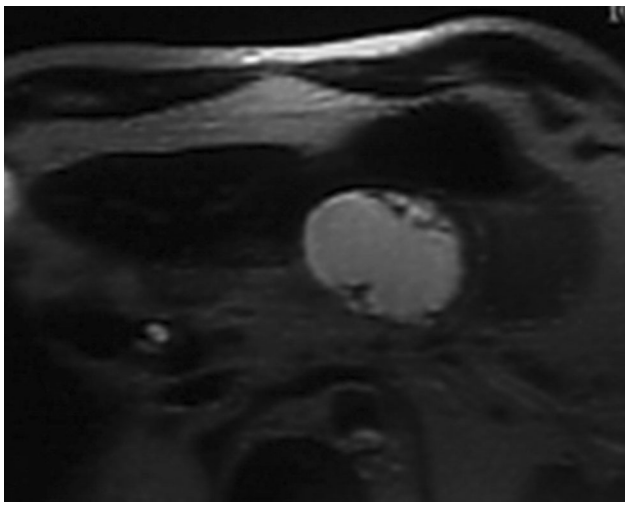

Fig. 2 T2-weighted magnetic resonance imaging of the tumor shows a hyperintense area occupying most of the tumor

Esophagogastroduodenoscopy (EGD) showed a submucosal tumor (SMT) with normal mucosa (Fig. 4). Because the intra-abdominal mass was suspected to represent a pancreatic cystic tumor based on the abdominal images, the findings from the EGD were assumed to represent an SMT-like finding with compression of the stomach wall by an extra-gastric pancreatic mass. However, endoscopic ultrasonography (EUS) revealed a cystic tumor located in the fourth layer of the stomach (Fig. 5). The area of cystic formation was fully filled with sludge-like material. Based on EUS findings, we finally diagnosed the intra-abdominal mass as a cystic tumor located in the gastric submucosa. As the patient declined to undergo EUS-guided biopsy because of the possibility of dissemination of the malignant cells into the intra-abdominal cavity, no pathological diagnosis could be reached.

Surgery was performed for the tumor. At the lesser curvature of the body of the stomach, a tumor covered on the smooth surface was detected. Neither metastasis of the lymph nodes nor invasion to the surrounding organs was detected. The tumor was not detected on the mucosa and serosa. The patient underwent subtotal gastrectomy and

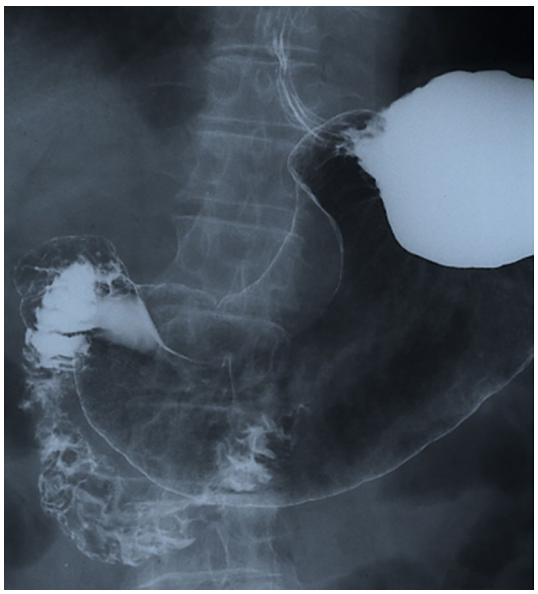

Fig. 3 Double-contrast gastrography shows the elevated lesion protruding into the gastric lumen at the lesser curvature of the body of the stomach, with a smooth surface

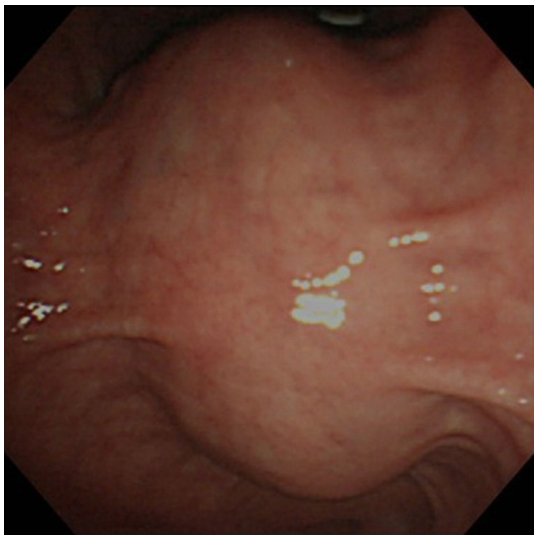

Fig. 4 Esophagogastroduodenoscopy shows the elevated lesion with a smooth surface

curative resection of the tumor was performed. The mass, measuring 60 × 50 × 50 mm, comprising both cystic and solid regions was resected (Fig. 6a). The cystic component made up the vast majority of the mass, and was filled by light bloody serous fluid. The tumor was located in the muscularis layer of the stomach (Fig. 6b). On histological examination, a solid region of the resected tumor showed a spindle-cell appearance (Fig. 7a) with positive staining for CD34 (Fig. 7b) and c-kit (Fig. 7c). The diagnosis was a gastric GIST with extreme predominance of the cystic component. The mitotic count was <5/50 high-power fields (HPFs) and the MIB-1 labeling index was <10 %.

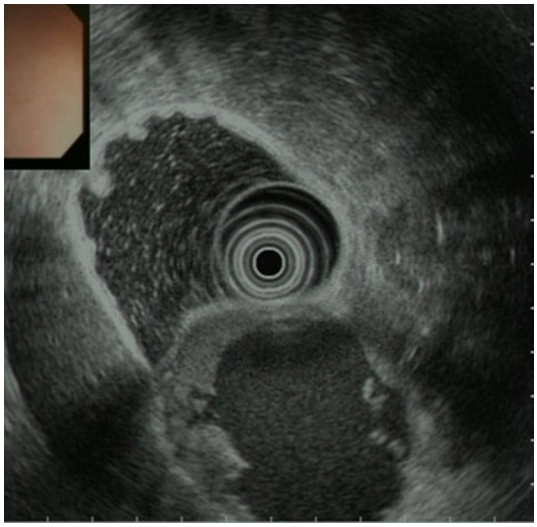

Fig. 5 Endoscopic ultrasonography reveals the cystic tumor located in the fourth layer of the stomach. The tumor was diagnosed as a submucosal tumor of the stomach

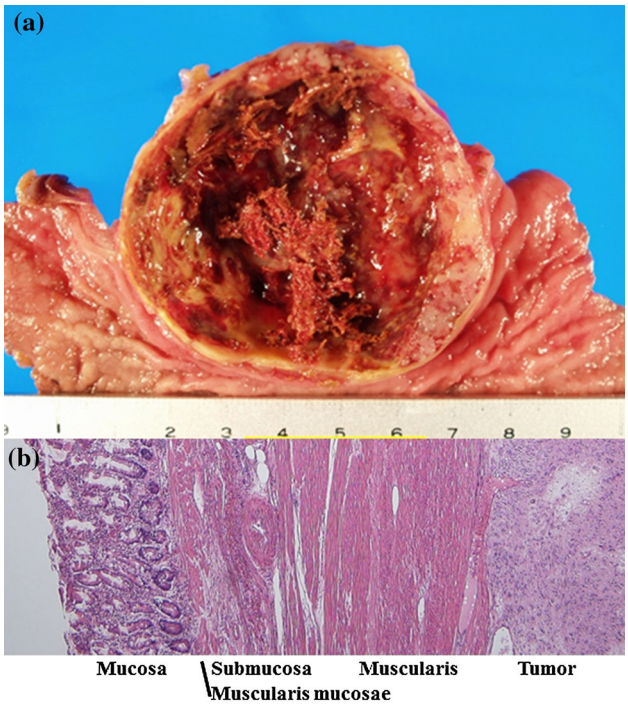

Fig. 6 The resected tumor shows components of both cystic and solid areas (**a**). The tumor located in the muscularis layer (**b**)

The GIST belonged to the intermediate-risk group based on Fletcher's classification system and Joensuu's classification system. However, the GIST belonged to the low-risk group based on Miettinen's classification system. Although the tumor was >50 mm in diameter, the patient did not undergo chemotherapy after the operation because we considered that the tumor had considerably less tumor cell volume than a whole solid tumor of the same size and its

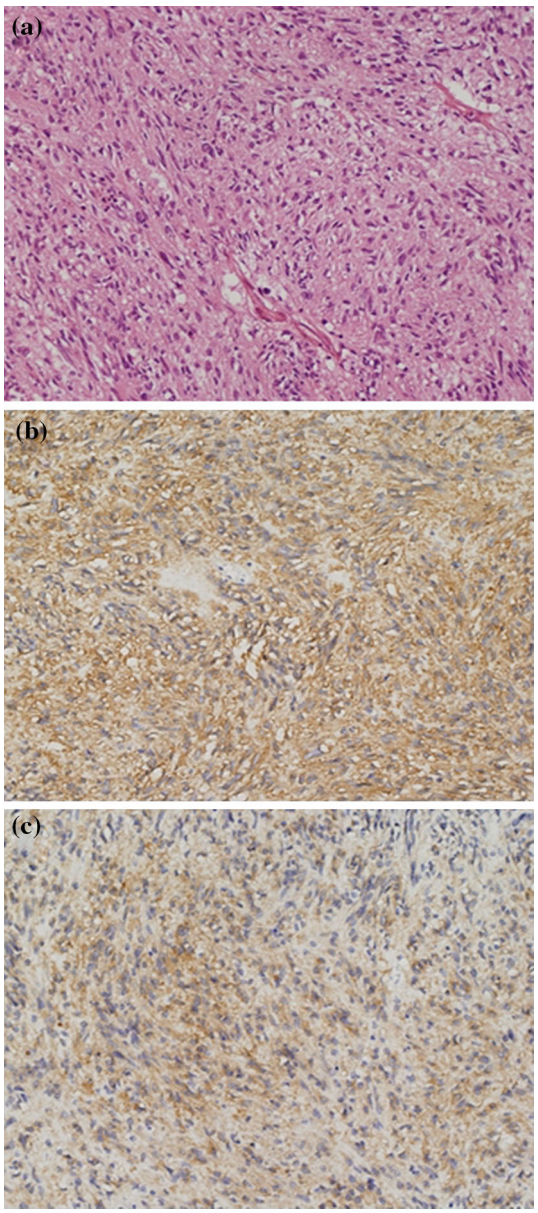

Fig. 7 Pathological findings reveal that the solid region of the resected tumor comprises spindle cells (**a**). The tumor shows positive staining for CD34 (**b**), and c-kit (**c**)

mitotic count and MIB-1 labeling index was low. Eventually the patient was observed with no treatment after surgery. Unfortunately, the patient died of arrhythmia 1 year postoperatively. No recurrence of the GIST was detected on autopsy.

Discussion

GISTs are usually solitary tumors, but the tumor in this case demonstrated predominant cystic formation. Reports of such cases of GISTs with predominant cystic changes are limited [1–14] and most seem to be misdiagnosed as deriving from liver or pancreatic tissue during preoperative diagnosis [11]. Although EUS is suggested as a useful modality for diagnosing GISTs with cystic changes [11], we could not decide whether this cystic mass represented a GIST or some other cystic tumor. EUS-guided biopsy, i.e., fine-needle aspiration (FNA) may be necessary to provide further pathological diagnostic evidence. The efficacy and accuracy of EUS-guided FNA in the diagnosis of GISTs has been previously reported [15]. However, in GISTs with predominant cystic formation, misdiagnosis might be more likely due to insufficient sample volume. EUS-guided FNA appears prudent for diagnosis of intra-abdominal cystic tumors, including GISTs. Transcutaneous aspiration biopsy for cystic tumors carries a risk of disseminating tumor cells into the intra-abdominal space.

In GISTs, a lesion with regions of hemorrhage or necrosis may form large cystic spaces [16], and GISTs often show extensive cystic changes in response to treatment with tyrosine kinase inhibitors [17]. However, our patient had not undergone any treatment before our diagnosis. The reason for the extensive cystic formation constituting the major portion of this GIST thus remains unclear. During tumor progression, massive necrotic changes of tumor cells might have occurred. Although most GISTs with cystic formation measure >10 cm in diameter [1–5, 8, 9, 11–14], the diameter of the tumor in our patient was 6 cm, and almost all of the tumor volume was occupied by a cystic lesion. Only one similar case of a stomach GIST lesion was found in the literature [7]. Our case indicates the possibility of giant cyst formation for relatively small GISTs. Small GISTs with cystic formation may be misdiagnosed, example, e.g., cystic pancreatic tumor.

When we face a predominantly cystic GIST, problems with determining tumor size may be encountered. At present, risk classification is recommended based on Fletcher [18], Miettinen [19] and Joensuu's [20] classification systems for GISTs. Fletcher's classification system is based on tumor size and mitotic count. Miettinen's classification system is based on tumor size, mitotic count and tumor site. Joensuu's classification system is a modified version of Fletcher's classification system and tumor rupture is proposed to be included in the high-risk category. Hence, tumor size is an important factor in tumor staging which influences the decision of whether to follow-up without treatment or perform resection. GISTs >5 cm in diameter are considered high-risk and might warrant additional chemotherapy after surgical resection [21]. However, in one case without imatinib treatment, no evidence of tumor recurrence or metastasis was recognized after resection of a >5-cm GIST with giant cystic formation [11]. In GISTs with predominantly cystic formation, the real tumor

volume of tumor cells is far smaller than the volume calculated as representing the tumor on imaging. A more accurate determination of the tumor cell volume of cystic GISTs may need to be computed by subtracting the cystic volume before deciding on the course of treatment. In the future, evaluation and follow-up of giant cystic GISTs may be necessary.

In conclusion, we encountered a rare case of a gastric GIST with predominant cystic formation occupying most of the tumor volume. This case suggests that we should keep GISTs in mind when encountering intra-abdominal cystic lesions. We should also be prudent in diagnosing predominantly cystic lesions in spite of relatively small-volume tumors.

Disclosures

Conflict of Interest: Hiroshi Okano, Tomomasa Tochio, Daisuke Suga, Hiroaki Kumazawa, Yoshiaki Isono, Hiroki Tanaka, Shimpei Matsusaki, Tomohiro Sase, Tomonori Saito, Katsumi Mukai, Akira Nishimura, Youichirou Baba and Tetsuya Murata declare that they have no conflict of interest.

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