Quiz

Correct answer to the quiz. Check your diagnosis

Case report

Synovial sarcoma of the stomach: case report and systematic review of the literature

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Synovial sarcoma is a rare mesenchymal malignant neoplasm that presents a specific t(X;18) translocation forming *SS18(SYT)-SSX* chimera gene. It is most commonly seen in soft tissues of the extremities. The digestive tract is an exceptional site of involvement. We report a case of primary gastric synovial sarcoma in a 48-year-old female. Differential diagnosis of synovial sarcoma from other spindle cell, mesenchymal and cytokeratin-positive tumors is critical for the treatment and prognosis. Immunohistochemistry studies and molecular analysis are required to settle a proper diagnosis.

Key words: synovial sarcoma, stomach, spindle cell neoplasm, cytokeratin.

Introduction

Synovial sarcoma is a rare mesenchymal malignant neoplasm, accounting for about 10% of soft tissue sarcomas. It is most commonly seen in soft tissues of the extremities, however, cases with unusual locations such as head and neck, lung and mediastinum, abdomen and retroperitoneum, kidney, among others, have been reported [1, 2]. Although the gastrointestinal tract is an extremely rare location for synovial sarcoma, this type of presentation has also been previously described and the stomach is the most frequent location along the digestive tract. To the best of our knowledge, only 36 primary gastric synovial sarcoma cases have been reported in the English literature [3-20]. We present an additional case of primary gastric synovial sarcoma in a 48-year-old female.

Case report

The patient was a 48-year-old female who presented at Specialist Hospital and Podkarpacki Oncology Center in Brzozow with nonspecific upper abdominal pain and intermittent nausea that had continued for a few months.

A CT scan of the abdominal cavity of the patient demonstrated extensive thickening of the wall of the pyloric region of the stomach (Fig. 1), but there was no other site indicating primary or metastatic lesions. A biopsy specimen, obtained during upper gastrointestinal endoscopy, revealed proliferation of atypical spindle cells positive for cytokeratin and negative for CD117, CD34 and SMA. According to the results, a GIST diagnosis was rejected and a suspicion of poorly differentiated carcinoma was made. Regarding the lesion size the patient received neoadjuvant chemotherapy. During surgery, a gastric tumor measured 9 × 8 × 3 cm was found. It involved the distal part of the body and the pyloric region of the stomach. The stomach was resected and both macroscopic and histological examination of the surgical specimen showed diffuse neoplastic infiltration covering the full thickness of the stomach wall (Fig. 2) and passing to the surrounding adipose tissue.

Material and methods

Immunostaining was performed for cytokeratins, EMA, Bcl2, SMA, Desmin, Vimentin, S100, CD31, CD34, CD117, CD99, SOX10 and HMB45. All immunohistochemistry was done on 4-μm-thick, standard-size sections, which were first dewaxed, rehydrated, and blocked for endogenous peroxidase (with 3.0% hydrogen peroxide). Details of primary antibodies, their dilutions, pretreatments, and sources used in the study are depicted in Table I.

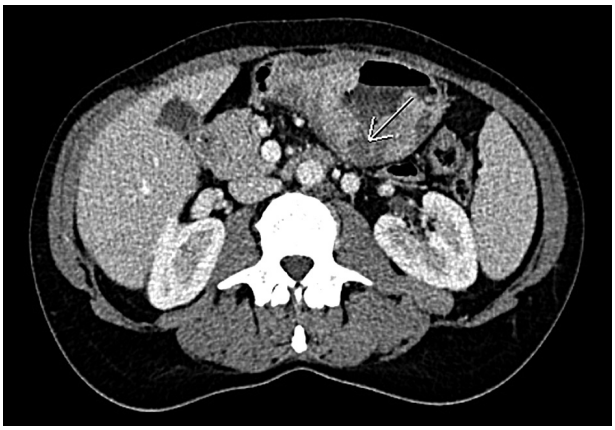

Fig. 1. CT scan of the abdominal cavity showing the location of the tumor

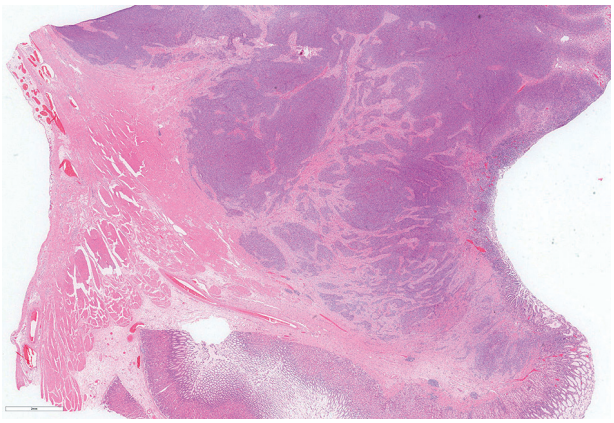

Fig. 2. The scan of the haemantoxylin and eosin (HE) staining slide covering the full cross-section of the stomach wall

Table I. Primary antibodies, their dilutions, pretreatments, and sources used in this study

Antigen	Antibody clone(s)	Dilution	Pretreatment	Source of antibody
Cytokeratins	AE1/AE3 cocktail	1:600	Citrate buffer – heat	Cell Marque
EMA	GP1.4	1:200	None	Leica Biosystem
Bcl2	124	1:100	Citrate buffer – heat	Cell Marque
SMA	OCSM-1	1:50	None	DAKO Cytomation
Desmin	DE-R-11	1:100	Citrate buffer – heat	Leica Biosystem
S100	polyclonal	1:600	Trypsin 15 min	Leica Biosystem
CD31	1A10	1:50	Citrate buffer – heat	Leica Biosystem
CD34	QBEnd/10	1:50	Trypsin 15 min	Leica Biosystem
CD117 (KIT)	A4502	1:400	Citrate buffer – heat	DAKO Cytomation
CD99	EPR30974	1:100	Citrate buffer – heat	Cell Marque
SOX10	N-20	1:100	Citrate buffer – heat	Santa Cruz Biotechnology
HMB45	HMB45	1:80	Citrate buffer – heat	Thermo Scientific
Vimentin	V9	1:250	Citrate buffer – heat	Cell Marque

Interphase fluorescent *in situ* hybridisation (FISH) was performed on 5 μm paraffin-embedded tissue sections using the LSI SS18 (18q11.2) Dual Color Break Apart Rearrangement Probe set (Vysis, Downers Grove, IL, USA). Hybridisation was performed according to the manufacturer's protocol. Slides were mounted and counterstained with anti-fade DAPI (Vysis, Downers Grove, IL, USA), visualized using an epifluorescent Microscope (Olympus BX61). At least 300 interphase nuclei were analyzed.

Results

Microscopically, HE slides showed monotonous histological texture built predominantly of spindle-shaped cells forming herring-bone or fibrosarcoma-like fascicles (Fig. 3). These spindle neoplastic cells were relatively small and uniform in size with fusiform or ovoid nuclei and a small or inconspicuous nucleoli. Their cytoplasm was scant and the cell borders were indistinct. Focally, the additional component made of epithelioid or small round cells have been also noticed (Fig. 4). Both spindle cells and the epithelioid or small round cells created focal hemangiopericytoma-like (Fig. 5) arrangements or formed solid sheets separated by fibrous or myxoid stroma (Fig. 6). Mitotic figures were readily identified with at least 7 figures per 10 high-power fields. The tumor stroma showed varying degrees of thin-walled dilated and/or staghorn-shaped blood vessels, thereby imparting a hemangiopericytoma-like growth pattern.

Immunohistochemically, the tumor cells were strongly positive for cytokeratins (AE1/AE3) (Fig. 7), EMA, vimentin (Fig. 8) and Bcl2 antigen (Fig. 9), but did not present reaction against smooth muscle actin, S100 protein, CD20, HMB45, SOX10, CD31, CD34, CD99 and CD117.

Rearrangement of the *SS18(SYT)* gene (18q11.2) was detected by dual-color break-apart fluorescent *in situ* hybridization analysis of the *SS18(SYT) locus* on an interphase cell nuclei (Fig. 10).

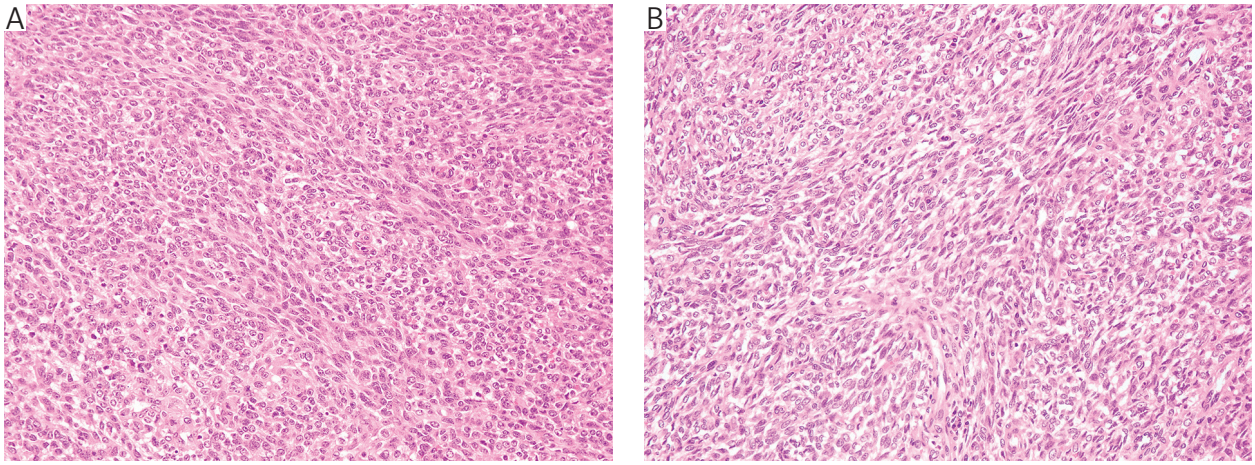

Fig. 3. A, B) Intersecting fascicles of the spindle cells in a herring-bone pattern (HE)

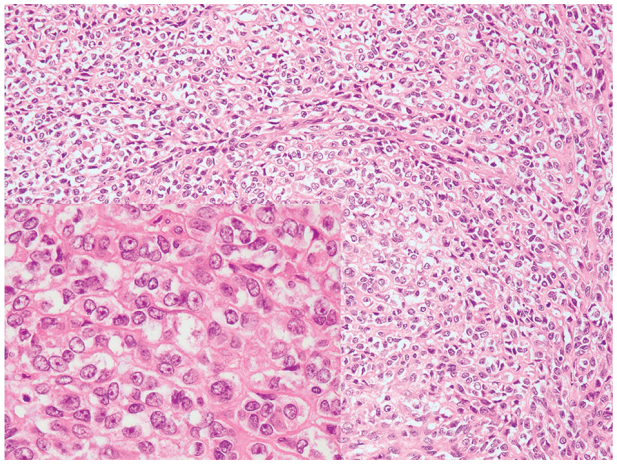

Fig. 4. Short spindle cell or epithelioid cell (insert) component of the tumor (HE)

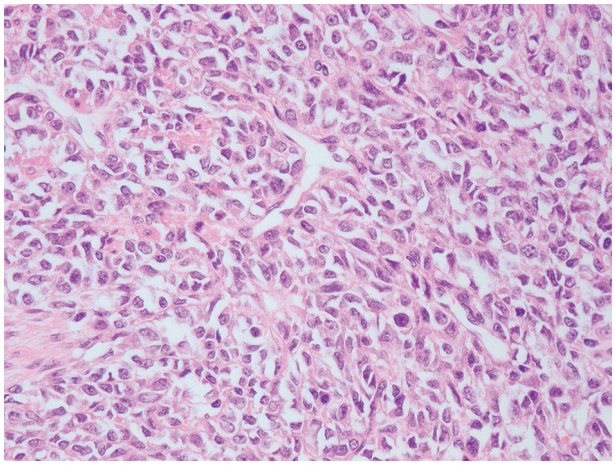

Fig. 5. Short spindle or small round cells creating hemangiopericytoma-like texture (HE)

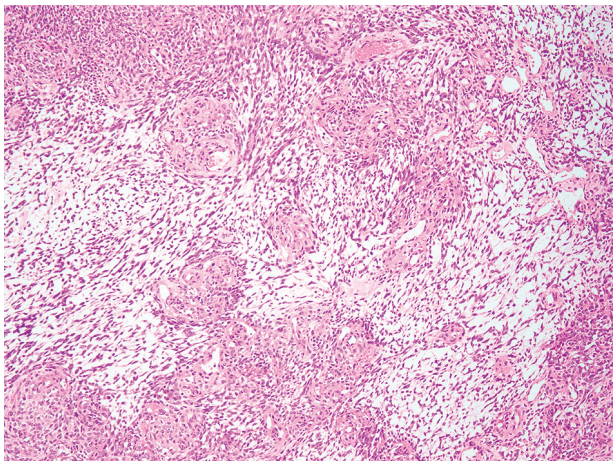

Fig. 6. Solid sheets of neoplastic cells separated by myxoid stroma (HE)

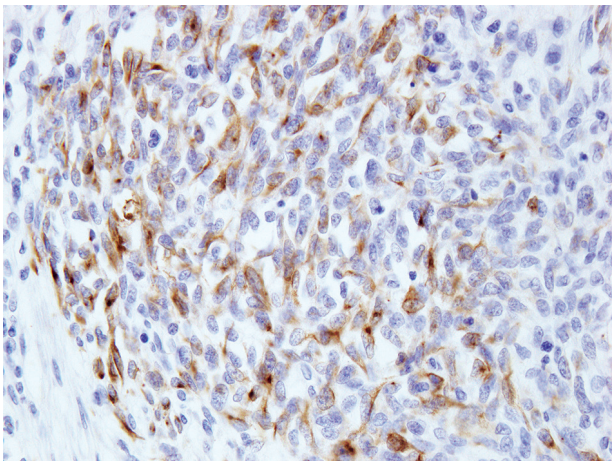

Fig. 7. Immunohistochemical staining for cytokeratins (AE1/AE3)

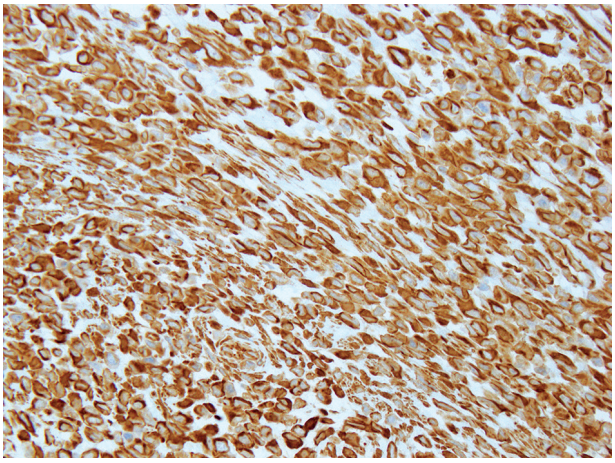

Fig. 8. Immunohistochemical staining for vimentin

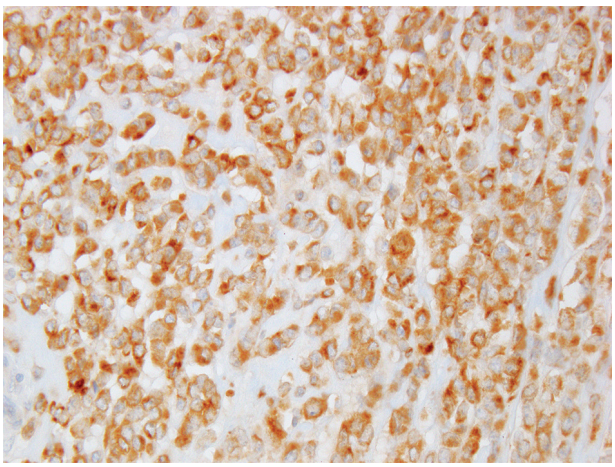

Fig. 9. Immunohistochemical staining for Bcl2

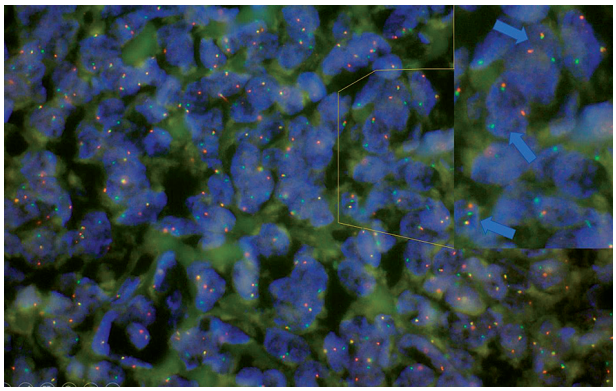

Fig. 10. FISH using *SS18(SYT)* break-apart probe set showed the split of two signals in most of the nuclei (original 1000× magnification)

Discussion

Synovial sarcoma (SS) can occur in many different locations throughout the body and is rarely found within the gastrointestinal tract. The first report concerning primary gastric SS was published in 2000 by Billings *et al*. [3]. The last two cases were reported by Manohar *et al*. [19] and Wong *et al.* [20] in 2020. The clinicopathological features of the 37 cases of primary gastric SS, including our case, are summarized in Table II.

Similarly to synovial sarcomas at other sites, gastric SS occur mostly in middle-aged patients with a median age of 47 years (range 13 to 68 years); there is a slight prevalence of males than females (19/18). The mean/median age of male and female patients is 40/42 (range from 13 to 62) and 51/50 (range from 35 to 68) years, respectively. The gastric body and fundus are the most common locations, but tumors localized in the gastroesophageal junction, cardia, antrum, and gastroduodenal junction have also been reported.

The tumor size ranged from 0.8 to 16 cm (median 4.35 cm). Most of the lesions were ulcerated. Ten tumors were transmural masses infiltrating adjoining

Table II. Clinico-morphological characteristics of the gastric synovial sarcoma (review)

No.	Gender	Age	Tumor size (mm)	Location	Neoplastic infiltration*	Histological subtype	Mitotic count (mitoses per HPF)	Confirmation	Fusion gene	Treatment	Follow-up (mo)	Outcome	Author, year, reference
1	M	47	52	Gastro-oesophageal junction	Not specified, pedunculated	Biphasic	1/10 HPF	FISH		Gastrectomy with partial esophagectomy	21	AND	Billings *et al*. Mod Pathol 2000 [3]
2	F	55	160	Distal stomach	Transmural, ulcerated	Biphasic / Poorly differentiated	9-50/10 HPF	FISH		Hemi-gastrectomy	6	DD	Billings *et al*. Mod Pathol 2000 [3]
3	M	42	115	Posterior gastric wall	Transmural	Biphasic	10/10 HPF	RT-PCR	*SS18(SYT)-SSX1*	Tumorectomy, chemotherapy	24	DD	Akhunji Cancer Ther 2007 [4]
4	F	67	8	Body-antrum junction	Mucosa, submucosa	Monophasic	0 - > 50 /10 HPF (in 4/10 cases > 15/10 HPF)	7×RT-PCR	3×*SS18(SYT)-SSX1* 4×*SS18(SYT)-SSX2*	Partial gastrectomy	12	AND	Makhlouf *et al*. Am J Surg Pathol 2008 [5]
5	M	49	20	Body	Mucosa, submucosa	Monophasic / Poorly differentiated				Wedge resection	29	DD	Makhlouf *et al*. Am J Surg Pathol 2008 [5]
6	F	68	20	Body	Mucosa, submucosa	Monophasic				Wedge resection	22	AND	Makhlouf *et al*. Am J Surg Pathol 2008 [5]
7	M	29	28	Body	Mucosa, submucosa	Monophasic				Partial gastrectomy	224	AND	Makhlouf *et al*. Am J Surg Pathol 2008 [5]
8	F	54	30	Antrum, gastroduodenal junction	Mucosa, submucosa + muscularis propria	Monophasic				Antrectomy/ gastroduodenal resection	ND		Makhlouf *et al*. Am J Surg Pathol 2008 [5]
9	F	58	30	Lesser curvature, body	Mucosa, submucosa + muscularis propria	Monophasic				Wedge resection	21	AND	Makhlouf *et al*. Am J Surg Pathol 2008 [5]
10	F	37	40	Fundus	Mucosa, submucosa + muscularis propria	Monophasic				Partial gastrectomy	48	Recurrence, died of other causes	Makhlouf *et al*. Am J Surg Pathol 2008 [5]
11	M	50	60	Distal fundus	Mucosa, submucosa + muscularis propria	Monophasic				Tumorectomy, chemotherapy	6	AD	Makhlouf *et al*. Am J Surg Pathol 2008 [5]
12	M	42	80	Greater curvature, body	Transmural	Biphasic				Partial gastrectomy, chemotherapy	25	DD	Makhlouf *et al*. Am J Surg Pathol 2008 [5]
13	F	66	150	Fundus	Transmural	Monophasic				Gastrectomy/ partial esophageectomy	Lost		Makhlouf *et al*. Am J Surg Pathol 2008 [5]

Table II. Cont.

No.	Gender	Age	Tumor size (mm)	Location	Neoplastic infiltration*	Histological subtype	Mitotic count (mitoses per HPF)	Confirmation	Fusion gene	Treatment	Follow-up (mo)	Outcome	Author, year, reference
14	F	44	47	Lesser curvature, body	Transmural + subserosal connective tissue	Monophasic	ND	FISH		Wedge resection	60	AND	Sinniah *et al*. Clin Transl Gastroenterol 2012 [6]
15	F	38	72	Body	Transmural +metastatsis in omentum	Monophasic	> 20/10 HPF	RT-PCR	*SS18(SYT)-SSX1*	Wedge resection, chemotherapy	6	AD	Wang *et al*. J Formos Med Assoc 2012 [7]
16	F	42	35	Body	Mucosa, submucosa, ulcerated	Monophasic	2/10 HPF	RT-PCR	*SS18(SYT)-SSX1*	Partial gastrectomy	72	AND	Kamata *et al*. Clin J Gastroenterol 2013 [8]
17	M	22	25	Body	Mucosa, submucosa + focally muscularis propia, ulcerated	Monophasic	14/10 HPF	RT-PCR	*SS18(SYT)-SSX1*	Wedge resection	ND		Sahara *et al*. Pathol Res Pract 2013 [9]
18	M	44	150	Lesser curvature, body	Transmural, ulcerated	Monophasic	11/10 HPF	FISH		Total gastrectomy	18	AND	Torres Rivas Pathology 2014 [10]
19	M	62	38	Fundus	Mucosa, submucosa	Monophasic	> 22/10 HPF	RT-PCR	*SS18(SYT)-SSX1*	Total gastrectomy, chemotherapy	9	AND	Michot *et al*. J Gastrointest Cancer 2014 [11]
20	F	50	80	Body	Mucosa, submucosa, muscularis propria	Monophasic	7/10 HPF	RT-PCR or both	*SS18(SYT)-SSX1*	ND	Lost		Romeo *et al*. Clin Sarcoma Res 2015 [12]
21	M	36	60	Cardias	Transmural + adventitia, peritoneum, omentum	Poorly differentiated	11/10 HPF	FISH		ND	36	AD	Romeo *et al*. Clin Sarcoma Res 2015 [12]
22	M	37	20	Gastric	Mucosa, submucosa, muscularis propria	Monophasic	6/10 HPF	FISH		ND	ND		Romeo *et al*. Clin Sarcoma Res 2015 [12]
23	M	26	NR	Gastric	Transmural + adventitia, peritoneum, pancreas	Monophasic	P	FISH		ND	185	AD	Romeo *et al*. Clin Sarcoma Res 2015 [12]

Table II. Cont.

No.	Gender	Age	Tumor size (mm)	Location	Neoplastic infiltration*	Histological subtype	Mitotic count (mitoses per HPF)	Confirmation	Fusion gene	Treatment	Follow-up (mo)	Outcome	Author, year, reference
24	M	58	100	Gastric	Transmural + adventitia, peritoneum, pancreas	Monophasic	12/10 HPF	RT-PCR or both	*SS18(SYT)-SSX1*	ND	6	DD	Romeo *et al*. Clin Sarcoma Res 2015 [12]
25	M	21	100	Gastric	Mucosa, submucosa, muscularis propria	Monophasic	P	FISH		ND	48	Lost	Romeo *et al*. Clin Sarcoma Res 2015 [12]
26	M	36	60	Gastric	Mucosa, submucosa, muscularis propria	Biphasic	27/10 HPF	RT-PCR or both	*SS18(SYT)-SSX2*	ND	12	Lost	Romeo *et al*. Clin Sarcoma Res 2015 [12]
27	F	54	38	Gastric	Mucosa, submucosa, muscularis propria	Monophasic	14	RT-PCR or both	*SS18(SYT)-SSX1*	ND	ND		Romeo *et al*. Clin Sarcoma Res 2015 [12]
28	F	49	35	Gastric	Submucosa, ulcerated	Monophasic	> 10/10 HPF	FISH		Tumorectomy	10	AND	Wong *et al*. Histopathology 2015 [13]
29	F	35	120	Gastric	Transmural + subserosal tissue, ulcerated	Monophasic	> 10/10 HPF	FISH		Tumorectomy, chemotherapy	48	AD	Wong *et al*. Histopathology 2015 [13]
30	F	51	9	Body	Submucosa	Monophasic	ND	RT-PCR	*SS18(SYT)-SSX1*	Distal gastrectomy	2	AND	So *et al*. Medicine (Baltimore) 2017 [14]
31	F	27	20	Gastric	Submucosa	Monophasic	ND	RT-PCR	*SS18(SYT)-SSX2*	Gastrectomy	6	AND	Ogino *et al*. World J Gastroenterol 2018 [15]
32	F	57	18	Lesser curvature, body	Submucosa, ulcerated	Monophasic	ND	RT-PCR	*SS18(SYT)-SSX2*	Wedge resection	ND	ND	Olsen *et al*. J Gastrointest Surg 2018 [16]
33	M	58	63	Greater curvature, body	Submucosa, ulcerated	Monophasic	33/10 HPF	FISH		Wedge resection	7	AD	Hu *et al*. J Gastrointest Cancer 2019 [17]
34	M	42	30	Lesser curvature, body	Not specified, pedunculated, ulcerated	Monophasic	3/25 HPF	RT-PCR	Not specified	Tumorectomy	12	AND	Fuente *et al*. J Gastrointest Surg 2019 [18]
35	M	13	110	Body, fundus	Submucosa, muscularis propria	Monophasic	Occasional	FISH		Total gastrectomy	6	AND	Manohar 2020 [19]

Table II. Cont.

No.	Gender	Age	Tumor size (mm)	Location	Neoplastic infiltration*	Histological subtype	Mitotic count (mitoses per HPF)	Confirmation	Fusion gene	Treatment	Follow-up (mo)	Outcome	Author, year, reference
36	M	54	16	Lesser curvature, body	Submucosa, muscularis propria, ulcerated	Monophasic	ND	FISH		Wedge resection	18	AND	Wong *et al.* Med J Hong Kong 2020 [20]
37	F	48	90	Body, pylorus	Transmural + adjacent adipose tissue	Monophasic	7/10 HPF	FISH		Distal gastrectomy, chemotherapy	Current case		This study 2020
Metastatic cases													
1	M	49	ND	ND	ND	Monophasic	ND	RT-PCR	*SS18(SYT)-SSX1*	Surgery, radiotherapy, chemotherapy	ND	ND	Anjiki *et al.* Gastrointest Endosc 2004 [22]
2	M	56	ND	ND	ND	ND	ND	ND	ND	Surgery, radiotherapy, chemotherapy	ND	AD	Samuel *et al.* Med J Malaysia 2016 [23]

*AND – alive with no evidence of disease; AD – alive with disease; DD – died of disease; P – pretreated; ND – no data; * – estimation based on accessible data; HPF – high-power fields*

tissues and the other twenty-seven were intramural and predominantly involving the 1umina1 side.

Histologically gastric SS presents similar microscopy characteristics to other locations. There are three classically defined microscopic variants: biphasic, monophasic and poorly differentiated. Among 37 tumors reported in the English literature (including our case) there were thirty cases of monophasic fibrous SS, four cases classified as biphasic subtype and other three cases with at least focal poorly differentiated component. Mitoses per 10 HPF ranged from 0 to over 50.

The immunohistochemical studies of this neoplasm have been varied, although determination of expression of wide spectrum cytokeratin was constant, obtaining positive results in all, except two reported cases (25/27). EMA, TLE1, CD56, Bcl2 and vimentin were positive in all tested cases. On the contrary, CD34, S100, SMA, and desmin were consistently negative (Tables III, IV). Almost all of the previously described gastric synovial sarcomas did not express CD117. The only exception were two cases presented by Wong *et al.* [13]. Two gastric synovial sarcomas from their collection showed a weak reaction to both CD117 and DOG1. However, no activating mutations were detected in *KIT* exons 9, 11, 13 and 17 or *PDGFRA* exons 12, 14 and 18 in any of these cases.

Therefore, a confident distinction between abdominal synovial sarcoma and GIST requires *KIT*/*PDGFRA* mutation analyses and specific molecular testing for synovial sarcoma. Most synovial sarcoma cases have a reciprocal translocation between the short arm of chromosome X and the long arm of chromosome 18. This translocation fuses the *SSX1* or *SSX2* genes from chromosome X and the *SS18(SYT)* gene from chromosome 18 to form *SS18(SYT)-SSX* chimera gene. The most reported gastric synovial sarcomas were molecularly confirmed by interphase FISH (14/37) or RT-PCR (20/37). Of 20 cases examined by RT-PCR at least two were simultaneously studied by FISH using *SS18(SYT)* break-apart probes [9, 12]. *SS18-SSX1* and *SS18-SSX2* fusion genes were demonstrated in 12 and 7 cases, respectively. In one case the fusion product was not specified [18].

Monophasic SS in the gastric wall should additionally be discerned from other mesenchymal tumors, including leiomyoma, leiomyosarcoma, schwannoma, solitary fibrous tumor, and "gastroblastoma" – a distinctive biphasic (epithelio-mesenchymal) tumor of the stomach in young adults [21], as well as other cytokeratin-positive tumors such as poorly differentiated carcinoma, and sarcomatoid carcinoma.

A diagnosis of synovial sarcoma should be considered particularly if an abdominal spindle cell neo-

Table III. Immunohistochemistry of gastric synovial sarcoma (review)

No.	References	CK (AE1/AE3)	EMA	VIM	CD99	TLE1	ChrA	SYN	CD117	S100	DES	SMA	HMB45	DOG1	CD34	CD56	Bcl2	CD57	CK7	Other
1	[3]	E+/S+	E+/S–	E–/S+	E–/S–	NP	NP	NP	NP	NP	NP	NP	NP	NP	NP	E–/S+	NP	E–/S–	NP	NP
2	[3]	E+/S+	E+/S-	E–/S+	E–/S-	NP	NP	NP	NP	NP	NP	NP	NP	NP	NP	E–/S+	NP	E+/S+	NP	NP
3	[4]	E+	E+	NP	S+	NP	–	–	–	NP	NP	–	–	NP	–	NP	NP	NP	E+	NSE(S+); LCA(–); TTF(–); MELAN-A(–)
4-11, 13	[5]	9+	9+	3+	1+	NP	NP	NP	8–	4–	4–	1+/3–	NP	NP	5–	1+	NP	NP	5+	NP
12	[5]	E+/S–	E+/S–	E–/S+	NP	NP	NP	NP	NP	NP	NP	NP	NP	NP	NP	NP	NP	NP	NP	NP
14	[6]	NP	NP	NP	+	NP	NP	NP	–	NP	NP	NP	NP	NP	NP	NP	NP	NP	+	NP
15	[7]	+	NP	NP	+	NP	NP	NP	–	NP	NP	–	NP	–	–	NP	NP	NP	-	WT-1(–); CD10(–); ER(–); PgR(–)
16	[8]	+	+	+	+	NP	NP	NP	–	–	–	–	NP	–	–	NP	+	NP	NP	NP
17	[9]	+	+	+	+	NP	NP	NP	–	–	–	NP	NP	NP	–	NP	+	NP	NP	NP
18	[10]	+	+	+	NP	NP	NP	NP	–	–	–	–	NP	–	–	NP	NP	NP	NP	NP
19	[11]	+	+	NP	NP	NP	NP	NP	NP	–	–	+	NP	+	–	NP	NP	NP	+	CK20(–); caldesmon(–)
20	[12]	NP	+	NP	+	+	NP	NP	–	–	–	–	NP	–	–	+	+	NP	NP	NP
21	[12]	NP	+	NP	+	+	NP	NP	–	–	–	–	NP	–	–	+	+	NP	NP	NP
22	[12]	+	+	NP	+	+	NP	NP	–	–	–	–	NP	–	–	+	+	NP	NP	NP
23	[12]	NP	+	NP	NP	NP	NP	NP	–	–	NP	NP	NP	NP	NP	NP	NP	NP	NP	NP
24	[12]	NP	+	NP	NP	NP	NP	NP	–	–	–	NP	NP	NP	–	NP	NP	NP	NP	NP
25	[12]	NP	+	NP	NP	NP	NP	NP	–	–	NP	NP	NP	NP	NP	NP	NP	NP	NP	NP
26	[12]	+	NP	NP	NP	NP	NP	NP	–	NP	NP	NP	NP	NP	–	NP	NP	NP	NP	NP
27	[12]	+	+	NP	+	+	NP	NP	–	–	–	–	NP	–	–	+	+	NP	NP	NP
28	[13]	–/+	+	NP	+	NP	–	–	+*	–	–	–	–	+*	+	+	NP	NP	NP	NP
29	[13]	–	+	NP	+	NP	–	–	+*	–	–	–	–	+*	–	+	NP	NP	NP	NP
30	[14]	+	NP	+	+	+	–	–	NP	–	–	NP	–	NP	NP	NP	NP	NP	NP	NP
31	[15]	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND
32	[16]	NP	NP	NP	NP	+	NP	NP	–	NP	NP	NP	NP	NP	NP	NP	NP	NP	NP	NP
33	[17]	+	+	NP	NP	+	NP	NP	–	–	–	NP	NP	–	–	NP	NP	NP	NP	NP
34	[18]	NP	+	NP	NP	NP	NP	NP	–	–	NP	NP	NP	NP	–	NP	+	NP	NP	AML(–); calretinin(+)

Table III. Cont.

No	References	CK (AE1/AE3)	EMA	VIM	CD99	TLE1	ChrA	SYN	CD117	S100	DES	SMA	HMB45	DOG1	CD34	CD56	Bcl2	CD57	CK7	Other
35	[19]	NP	+	NP	NP	+	NP	NP	–	–	–	–	–	–	–	NP	+	NP	NP	STAT6(–); β-catenin(–); myogenin(–); Myo D1(–); ALK(–)
36	[20]	–	NP	NP	NP	+	NP	–	–	–	–	–	–	–	–	NP	NP	NP	NP	STAT6(–);ALK(–); calretinin(–); CD31(–); CD45(–); CAM5.2(–)
37		+	+	+	–	NP	NP	NP	–	–	NP	–	–	NP	–	NP	+	NP	NP	CK20(–); SOX10(–); CD31(–)
Metastatic cases																				
1	[22]	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND
2	[23]	+	+	+	–	NP	NP	NP	–	NP	–	–	NP	–	–	NP	+	NP	NP	NP

*E – epithelial cells; S – spindle cells; NP – not performed; ND – no data; * – weak intensity; –/+ < 10% neoplastic cells immunopositive*

Table IV. Summary of immunohistochemistry results

	CK (AE1/AE3)	CK7	EMA	VIM	CD99	TLE1	Bcl2	CD56	CD34	CD117	DOG1	S100	DES	SMA	ChrA	SYN
+	25	8	30	11	13	9	9	9	1	2*	3 (2*)	0	0	2	0	0
–	2	1	0	0	3	0	0	0	23	28	10	23	19	16	4	5
NR	10	28	7	26	21	28	28	28	8	7	24	14	18	19	33	32

*NR – not reported; () – including; * – weak intensity*

Table V. Synovial sarcoma of the stomach. Disease specific survival depending on the histological type of the tumor

Histological subtype	Alive	Died	Total
Monophasic	19 (95%)	1 (5%)	20
Biphasic	1 (33.3%)	2 (66.67%)	3
Poorly differentiated	1 (33.3%)	2 (66.67%)	3
All	**21 (80.77%)**	**5 (19.23%)**	**26**

χ^2 Pearson's coefficient	**p = 0.00352**
χ^2 NW	**p = 0.00716**

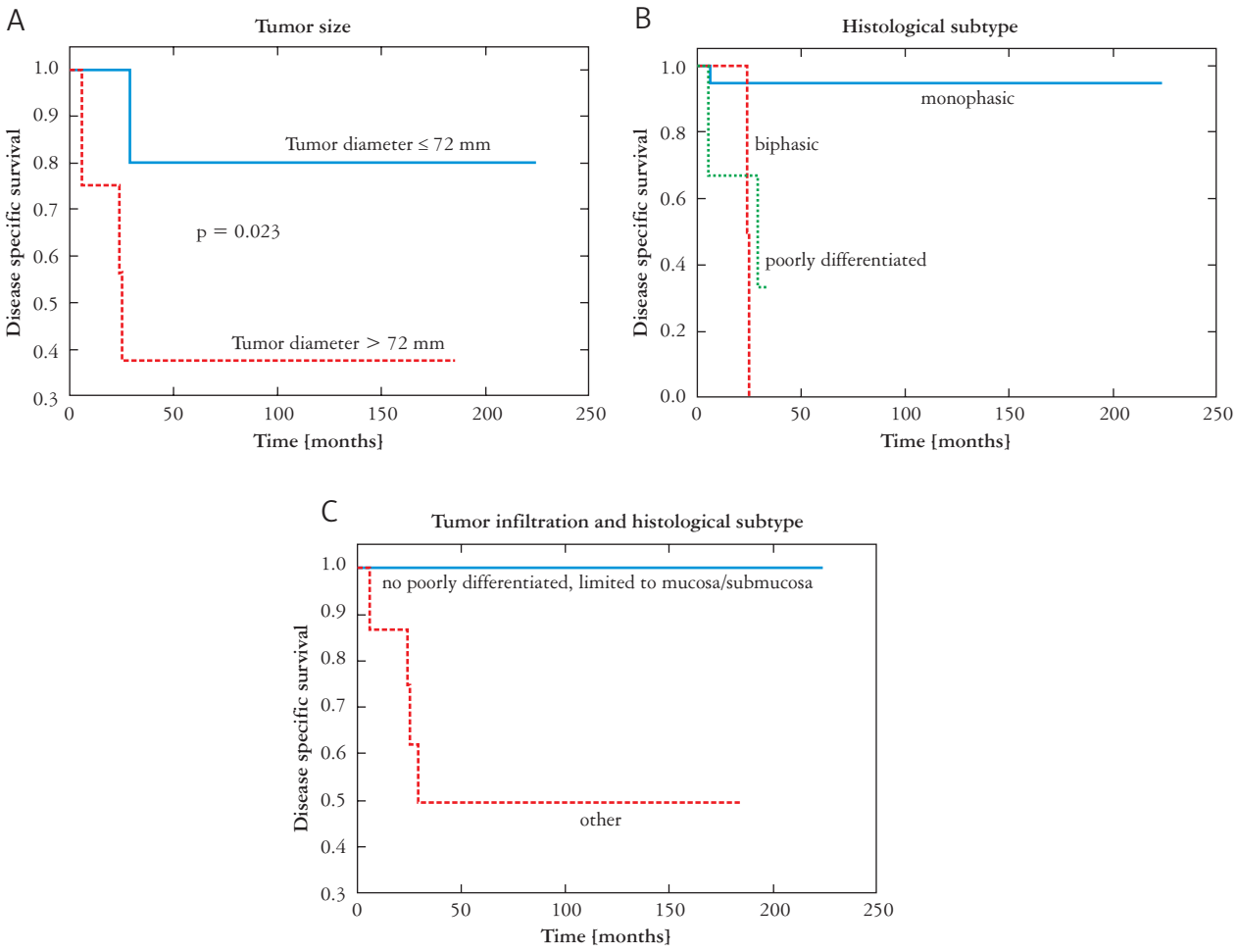

Fig. 11. Three morphological features affecting disease-specific survival of the gastric synovial sarcomas: A) tumor size, B) histological subtype of tumor, and C) depth of infiltration of the stomach wall together with a histological subtype of the tumor

plasm shows a haemangiopericytoma-like pattern and diffuse CD99 immunopositivity.

The mainstay of treatment for gastric SS is surgery, such as total or partial gastrectomy and wedge resection. All the reported cases, including our case, have undergone surgical resection. One (our) case has received neoadjuvant chemotherapy and other six cases have received postoperative chemotherapy; however, none of them has received radiotherapy.

Prognosis of SS in the gastrointestinal tract is unclear because of too small number of cases. The reviewed publications document six deaths, from which five were directly related to this disease. Two cases were defined as biphasic (tumor size 8 and 11.5 cm), one belonged to monophasic subtype (10 cm) and

other two had poorly differentiated component (one biphasic – 16 cm; one monophasic – 2 cm). Those deaths occurred between 6 and 29 months following diagnosis. One patient, with local recurrence, died of causes not related to the SS.

Survival analysis was carried out in the group of 26 patients for whom data concerning outcome and follow up were available. In this group 5 deaths were noted and all were caused by this disease. The median time of observation was 18 months and ranged from 2-224 months.

The mean tumor size in the group of patients who survived was statistically smaller that in the group of patients who died of disease (52.6 mm vs. 95 mm, $p = 0.049$). We found out that patients with tumor larger than 72 mm had statistically significantly worse probability of survival ($p = 0.023$) (Fig. 11A).

It seems that histological subtype can influence the prognosis. In the group of patients with monophasic subtype only one patient died (1/20), whereas in the group with biphasic or poorly differentiated tumors the percentage of deaths was significantly higher (Table V, Pearson χ^2 $p = 0{,}004$). These observations were confirmed by Kaplan-Meier analysis of survival (Fig. 11B).

Although there is no statistical significance, it was observed that in the group of patients who had tumor limited to mucosa or/and submucosa and without poorly differentiated component, the survival was 100% ($p = 0.143$) (Fig. 11C).

This shows that the tumor size and histological subtype of SS arising in the stomach are important prognostic factors.

Metastatic synovial sarcoma has been reported at least twice in the gastric wall (Table II). This was a case of a 49-year-old Japanese man who underwent chemotherapy and radiotherapy after amputation of his right leg for monophasic fibrous synovial sarcoma of the right calf. He developed multiple metastatic lesions in the stomach and duodenum; histologically similar to the primary tumor [22]. The second case was a 56-year-old male with primary synovial sarcoma of the left thigh with subsequent metastases to the stomach [23].

Conclusions

In summary, the primary gastric SS is a rare and underdiagnosed neoplasm. The awareness of occurrence of synovial sarcoma in the stomach may help to settle the proper diagnosis of tumor that should not be confused with other spindle cell and cytokeratin-positive neoplasms, as well as KIT-negative GISTs.

The authors declare no conflict of interest.

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