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 $9 \times 8 \times 3$ 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.

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).
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...
| No. | Gender | Age | Tumor size (mm) | Location | Nodular infiltration | Histological subtype | Mitotic count (mitoses per HPF) | Confirmation | Fusion gene | Treatment | Follow-up (mo) | Outcome | Author, Year, Reference |
|-----|--------|-----|----------------|----------|---------------------|---------------------|-------------------------|-------------|------------|-----------|-------------|---------|------------------------|
| 1   | M      | 47  | 52             | Gastroesophageal 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 | Partial gastrectomy | 12       | AND | Makhlouf et al. Am J Surg Pathol 2008 [5] |
| 5   | M      | 49  | 20             | Body | Mucosa, submucosa | Monophasic | Poorly differentiated |           | 4×SS18(SYT)-SSX2 | Partial gastrectomy | 29       | DD | Makhlouf et al. Am J Surg Pathol 2008 [5] |
| 6   | F      | 68  | 20             | Body | Mucosa, submucosa | Monophasic |           |           |           | Partial gastrectomy | 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 |           |           |           | Antrectomy/gastroduodenal resection | ND       |           | 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 esophagectomy | Lost     |           | Makhlouf et al. Am J Surg Pathol 2008 [5] |
| No. | Gender | Age (yr) | 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 | SS18(SYT)-SSX1 | Wedge resection | 60 | AND | Sinniah et al. Clin Transl Gastroenterol 2012 [6] |
| 15  | F      | 38       | 72              | Body | Transmural + metastasis 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 propria, 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 | ND | | 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 (yr) | 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         | SS18(SYT)-SSX1 | 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         | SS18(SYT)-SSX1 | ND       | 10        | AND      | Wong et al. Histopathology 2015 [13] |
| 29  | F       | 35       | 120             | Gastric  | Transmural + subserosal tissue, ulcerated | Monophasic           | > 10/10 HPF | FISH         | SS18(SYT)-SSX2 | ND       | 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         | SS18(SYT)-SSX2 | 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       | SS18(SYT)-SSX2 | Wedge resection | 12 | AND | Fuente et al. J Gastrointest Surg 2019 [18] |
| 35  | M       | 13       | 110             | Body, fundus | Submucosa, muscularis propria | Monophasic           | Occasional      | FISH         | SS18(SYT)-SSX2 | Total gastrectomy | 6 | AND | Manohar 2020 [19] |
| No. | Gender | Age | Tumor Size | Location | Neoplastic Infiltration | Histological Subtype | Mitotic Count (per HPF) | Confirmation | FUSION GENE | Treatment | Follow-up (mo) | Outcome | Treatment Follow-up | Outcome Author, Year, Reference |
|-----|--------|-----|------------|----------|------------------------|----------------------|------------------------|--------------|-------------|------------|--------------|----------|---------------------|----------------------------------|
| 36  | M      | 54  | 16         | Lesser curvature, body | Monophasic fibrous | 7/10 HPF             | ND                     | FISH         | SS18(SYT)-SSX1 | Wedge resection | 18          | AND                  | Died, gastroscopy, chemotherapy | Wong et al., Med J Hong Kong 2020 [20] |
| 37  | F      | 48  | 90         | Body, pylorus | Monophasic fibrous, ulcerated | Monophasic            | ND                     | FISH         | SS18(SYT)-SSX1 | Distal gastrectomy, chemotherapy | Current case | ND                  | ND                  | Wong et al., Med J Hong Kong 2020 [20] |

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

- 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/PDGFR 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 | Ckit| SYN | CD117 | S100 | DES | SMA | HMB-45 | DOG1 | CD34 | CD56 | Bcl2 | CD57 | CK7 | Other |
|-----|------------|--------------|-----|-----|------|------|-----|-----|------|------|-----|-----|--------|------|------|------|------|------|-----|-------|
| 1   | [3]        | E+/S+        | E+/S- | E-/S- | NP   | NP   | NP  | NP  | NP   | NP   | NP  | NP  | NP     | NP   | NP   | NP   | NP   | NP   | NP  |      |
| 2   | [3]        | E+/S+        | E+/S- | E-/S- | NP   | NP   | NP  | NP  | NP   | NP   | NP  | NP  | NP     | NP   | NP   | NP   | NP   | NP   | NP  |      |
| 3   | [4]        | E+           | E+   | NP   | S+   | NP   | –   | –   | NP   | –    | –   | NP   | NP     | NP   | NP   | E+   |      |      |     |      |
| 4-11, 13 | [5]   | 9+           | 9+   | 3+   | 1+   | NP   | NP   | NP  | 8-   | 4-   | 4-   | 1+/3- | NP   | NP   | 5-   | 1+   | NP   | NP   | 5+   |     |      |
| 12  | [5]        | E+/S-        | E+/S- | E-/S+ | 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   | +    |      |      |      |
| 15  | [7]        | +            | NP   | NP   | +    | NP   | NP  | NP  | NP   | –    | NP  | NP  | NP     | NP   | NP   | NP   | NP   |      |      |      |
| 16  | [8]        | +            | +    | +    | +    | NP   | 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   | 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     | –    | +    | +    | NP   | NP   | NP  |      |
| 21  | [12]       | NP           | NP   | NP   | +    | NP   | NP  | NP  | –    | –    | –    | NP   | +    | NP     | +    | NP   | NP   | NP   | NP   | NP  |      |
| 22  | [12]       | +            | +    | NP   | +    | NP   | NP  | NP  | –    | –    | –    | 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   | NP  |      |
| 24  | [12]       | NP           | NP   | NP   | 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   | NP  |      |
| 26  | [12]       | NP           | 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   | –    | NP  | NP  | NP     | NP   | NP   | NP   | NP   | NP   | NP  |      |
| 28  | [13]       | –/+          | +    | NP   | NP   | NP   | –    | –    | +*   | –    | –    | –    | –    | +*     | –    | –    | –    | +    | NP   | NP  |      |
| 29  | [13]       | –            | +    | NP   | NP   | NP   | +    | +*   | –    | –    | –    | –    | +*     | –    | +    | NP   | NP   | NP   | NP  |      |
| 30  | [14]       | +            | NP   | NP   | NP   | +    | NP   | –    | 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  |      |
| 32  | [16]       | 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   | NP   | NP   | NP  |      |
| 34  | [18]       | NP           | NP   | NP   | NP   | NP   | NP   | NP  | –    | NP   | +    | NP  | NP     | NP   | NP   | AML(−); calretinin(−) |
Table III. Cont.

| No | References (AE1/AE3) | CK (AE1/AE3) | EMA | VIM | CD99 | TLE1 | ChiRA | SYN | CD117 | S100 | DES | SMA | HMB-45 | 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  | NP   | STAT6(–); ALK(–); calretinin(–); CD31(–); CD45(–); CAM5.2(–) |
| 37 |                      | +           | +   | +   | –    | NP   | NP    | NP  | –     | –    | NP  | –   | NP     | –    | NP   | +    | NP   | NP   | NP  | CK20(–); SOX10(–); CD51(–) |

Metastatic cases

|        | ND | ND | ND | ND | ND | ND | ND | ND | ND | ND | ND | ND | ND | ND | ND | ND | ND | ND | ND |
|--------|----|----|----|----|----|----|----|----|----|----|----|----|----|----|----|----|----|----|----|
| 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  | NP  | ND |

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 | ChiRA | SYN |
|---------|--------------|-----|-----|-----|------|------|------|------|------|-------|------|------|-----|-----|--------|-----|
| +       | 25           | 8   | 30  | 11  | 13   | 9    | 9    | 1    | 2*   | 3 (2*)| 0    | 0    | 2   | 0   | 0      |     |
| –       | 2            | 1   | 0   | 0   | 0    | 0    | 0    | 3    | 28   | 23    | 28   | 10   | 23  | 19   | 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
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

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**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 |

χ² Pearson’s coefficient, p = 0.00352
χ² NW, p = 0.00716

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**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
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 χ² 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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