Long-term disease-free survival of an undifferentiated pleomorphic sarcoma of the spleen
A case report and literature review

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Abstract

Introduction: Undifferentiated pleomorphic sarcoma (UPS) primarily occurs in the soft tissues of the extremities, trunk, and retroperitoneum. As the primary UPS of the spleen (splenic UPS) is extremely rare, to the best of our knowledge, only 19 cases have been reported in English literature. No cases of long-term survival without a local or distant recurrence have been reported.

Patient concerns: We report the case of a 37-year-old man who was referred to our hospital for a splenic tumor. He had no past medical or relevant familial history. On abdominal computed tomography (CT), a low attenuation solid mass and cystic component with mural calcifications were present at the lower pole of his spleen. The fluorodeoxyglucose-positron emission tomography (CT) indicated it as malignant tumor of the spleen.

Diagnoses: The patient’s provisional diagnosis was deduced to be angiosarcoma, which was the most common malignant tumor of the spleen.

Interventions: An elective laparoscopic splenectomy was performed, and the histology of the tumor was consistent with UPS (pT1, pN0, cM0, and AJCC8th). No adjuvant therapy was administered.

Outcomes: Ten years have passed since the patient’s splenectomy, and he continues to do well, without evidence of local or distant recurrence.

Lessons: To the best of our knowledge, this is the first case of long-term recurrence-free survival after surgical management of a splenic UPS. It is probable that radical splenectomy during the disease played the most important role in the patient’s long-term survival. Understanding the characteristic findings of a splenic UPS in an abdominal CT may help to diagnose properly.

Abbreviations: AJCC = American Joint Committee on Cancer, CT = computed tomography, MFH = malignant fibrous histiocytoma, UPS = undifferentiated pleomorphic sarcoma.

Keywords: case report, long-term survival, MFH, spleen, UPS

1. Introduction

Primary undifferentiated pleomorphic sarcoma (UPS) is the most prevalent type of soft tissue sarcoma. It is usually encountered in the extremities and sometimes in retroperitoneum. The 5-year overall survival rate is 42% to 60%. However, UPS originated in the visceral organs is extremely rare and only a limited number of cases have been reported. Due to lack of appropriate follow-up data, its prognosis has still been unsure. UPS of spleen origin (splenic UPS) is also extremely rare and long-term survival case has never been reported. Herein, we report the case of a patient with a splenic UPS who is alive without a local or distant recurrence over 10 years after surgery, and survey of the literature for important points on this disease.

2. Case report

A 37-year-old man was referred for evaluation of a splenic tumor that was detected on ultrasonography at an outside hospital a month before during a workup for chest pain. Although the cause of the patient’s chest pain remained unclear, it was self-limited. He did not have any past medical history, any drug...
allergy or relevant familial history. He had no history of alcohol intake and smoking. His abdomen was soft, nondistended, and nontender, without a palpable mass. His vital signs were normal. Clinical laboratory data were unremarkable: white blood cell count $5.49 \times 10^3/\mu L$, hemoglobin $15.8\, g/dL$, platelet count $232 \times 10^3/\mu L$, LDH $141 U/L$, CRP $0.25\, mg/dL$, CEA

**Figure 1.** Preoperative CT and FDG-PET CT picture. (A) Computed tomography (CT) demonstrated a low attenuation solid mass protruding from the lower pole of the spleen (white arrow). (B) A cystic component with mural calcifications adjacent to the mass was shown. (arrow head). (C and D) The tumor was FDG avid on FDG-PET/CT. FDG-PET/CT = fluorodeoxyglucose-positron emission tomography.

**Figure 2.** Specimen examination. (A and B) The tumor size was $5 \times 5 \times 4.5\, cm$ and the large cystic component of the tumor contained necrotic and degenerative tissue. The tumor did not expose to splenic capsule and the surgical margin was negative. (C and D) Histologic examination of resected tissue samples. Hematoxylin and eosin (H&E) staining, original magnification ×40 (C), ×200 (D). It showed spindle-shaped cells arranged in a storiform pattern and accompanied by fibrous tissue.
### Table 1
Summary of reported cases of splenic undifferentiated pleomorphic sarcoma.

| Case no | Literature studies | Age/Gender | Tumor size (cm) | T and M value (AJCC 8th) | Synchronous metastasis | Clinical manifestation | CT findings | IHC staining (positive) | Treatment | Survival (after surgery) |
|---------|--------------------|------------|-----------------|--------------------------|------------------------|-----------------------|-------------|------------------------|-----------|-------------------------|
| 1       | 1982, Govoni[12]    | 51/F       | 21 × 25 × 10    | N/A                      | None                   | Abdominal pain, weight loss | N/A         | N/A                    | Splenectomy | Alive at 7 months       |
| 2       | 1982, Wick[13]      | 54/M       | N/A             | N/A                      | N/A                    | None                  | N/A         | N/A                    | N/A        | Alive at 3 months       |
| 3       |                    | 48/M       | 8               | M1                       | Liver                  | N/A                   | N/A         | N/A                    | N/A        | Alive at 18 months      |
| 4       | 1988, Bruneton[14]  | 51/F       | N/A             | T1 (stomach, pancreas)   | None                   | N/A                   | N/A         | N/A                    | Splenectomy | Alive at 17 months      |
| 5       | 1990, Siebert[15]   | 41/M       | N/A             | M1                       | Omentum, Peritoneum    | N/A                   | Cystic component Calcification | N/A        | Splenectomy             | Died at 6 months         |
| 6       | 1993, Lieu[16]      | 71/M       | N/A             | M1                       | Liver                  | N/A                   | N/A         | N/A                    | Vimentin, CD68 | Died after surgery       |
| 7       | 1994, Bornilla[17]  | 42/F       | N/A             | M1                       | Bone marrow            | N/A                   | N/A         | N/A                    | Splenectomy | Died at 8 months         |
| 8       | 1998, Mallpudi[18]  | 73/M       | 10              | T3 (retroperitoneum)     | None                   | Fever, Night sweat Weight loss | N/A         | N/A                    | Splenectomy | Died at 18th months      |
| 9       | 2001, Colovic[19]   | 45/F       | 11 × 10 × 7     | T1/T2                   | T1/T2                  | Abdominal pain, Weight loss Fever, Night sweat | N/A         | Vimentin, CD68, HLA-DR, lysozyme, S-100 | Splenectomy | Died at 15 months       |
| 10      | 2003, Gaaraas[20]   | 51/F       | 12 × 11 × 10    | T1/2                     | None                   | Abdominal pain, Weight loss Fever, Night sweat | N/A         | Cystic component Mural calcification | Splenectomy | Not written              |
| 11      | 2006, Katsura[21]   | 82/M       | 2.5 × 3         | T2                       | None                   | Abdominal pain, Weight loss Fever | Low density mass | N/A                    | Splenectomy | Alive at 18 months      |
| 12      | 2010, Hashmi[22]    | 76/M       | 7.1 × 5.3       | T1/T2                   | None                   | Abdominal pain         | Cystic component | N/A                    | Splenectomy | Not reported             |
| 13      | 2011, He[23]        | 35/M       | 5 × 5           | T1/T2 (rupture)          | None                   | Abdominal pain         | N/A         | Vimentin, αSMA, α1-antichymotrypsin | Splenectomy | Died at 7 months         |
| 14      | 2011, Ji-Feng[24]   | 48/M       | 5.2 × 4.6       | T1/T2                   | None                   | Abdominal pain         | Low density mass     | Vimentin, CD68, α1-antichymotrypsin | Splenectomy | Alive at 13 months      |
| 15      | 2011, Amatya BM[25] | 77/M       | N/A             | T3 M1 (rupture)          | Renal hilum Adrenal grand Femoral bone marrow | Hemorrhagic shock | N/A         | Vimentin, CD68 | Splenectomy | Died without surgery     |
| 16      | 2012, Dawsonson[26] | 30/M       | N/A             | T1/T2                   | None                   | Abdominal pain         | Solid mass Cystic component | N/A        | Vimentin, CD68 | Not followed |
| 17      | 2017, Makis[27]     | 63/M       | 7.5 × 7.3 × 7   | M1                      | Liver                  | Fever, Night sweat     | Solid mass Peripheral enhancement | EBER, Fascin | Splenectomy | Died at 16 months without surgery |
| 18      | 2020, Ashmore[28]   | 56/F       | 15              | T3 (diaphragm)          | None                   | Abdominal pain, anemia | Cystic component Mural calcification | CD31 | Splenectomy | Died at few months       |
| 19      | 2022, Tomioka[29]   | 37/M       | 5 × 5 × 4.5     | T1                      | None                   | Chest pain             | Solid mass Cystic component Mural calcification | Vimentin, CD68, α1-antichymotrypsin | Lap-Splenectomy | Alive at 10 years       |

IHC = immunohistochemistry, N/A = information not available.
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The purpose of adjuvant radiotherapy is to inactivate the microscopic extensions of tumor and histologically positive margins. In the case, the tumor hadn’t extended beyond the splenic capsule, and pathological R0 resection was performed. This was the reason why adjuvant radiotherapy was not performed. In 15 cases, including this case, a splenectomy was performed and a laparoscopic splenectomy was reported in 2 cases. In 2 cases, surgical treatment was not selected, because the diseases were too progressed. In the other 3 cases, we could not find descriptions about the way of treatment. Although the actual recurrence-free survival of surgically managed splenic UPS has not been reported, the longest individual recurrence-free survival of the cases currently available in the literature was 18 months. More than 10 years of long-term postoperative survival has never been reported. It may indicate that splenic UPS has an aggressive malignancy with a high potential for local recurrence and distant metastases. We reviewed all reported cases of splenic UPS to measure the primary tumor (T) category of each tumor if possible using the American Joint Committee on Cancer (AJCC) 8th Edition (Table 2).[9,10] Distinct metastases at presentation were classified as M1. Prognostic staging has never been defined because there are limited data on UPS of the peritoneal cavity, including the spleen. We could evaluate the tumor status in 16 of 20 cases (80%). Three patients were T3 or T4 and M0 (18.8%) and 6 were M1 (37.5%); in 2 cases (No.14 and No.16), the tumor was ruptured (12.5%). Among them, 9 patients died within 19 months of surgery or diagnosis and 1 patient was alive with liver metastases 18 months after surgery (Case No. 3). The other 6 cases (37.5%) that were T1 or T2 and M0 were alive without recurrence at the time that their cases were published. These results may indicate that the reasons why splenic UPS is thought to have an extremely poor prognosis are that the majority of patients with splenic UPS have evidence of adjacent organ invasion or distant metastasis or that the tumor is ruptured when it is discovered. Conversely, if a splenic UPS is discovered early during the disease and resected radically, long-term survival might be expected. Unfortunately, in most cases, splenic tumors

3. Discussion

This case is, to our knowledge, the first of a long-term recurrence-free survival after a splenectomy for a splenic UPS. UPS, previously known as malignant fibrous histiocytoma (MFH), represents a heterogeneous group of sarcomas without a specific known line of differentiation. It is most frequently encountered as a malignant soft tissue tumor of the extremities, and primary UPS of the spleen is extremely rare.[1,2] The first reported case of splenic UPS was termed a splenic MFH by Govoni in 1982. Only 19 total cases have been reported in English literature.[12-28] Details of previous studies on splenic UPS, including the present work, are shown (Table 1). Fourteen of the (70.0%) patients were male, and 6 (30.0%) were female. The mean age was 54.3 years (range: 30–82 years). The most common symptoms of a splenic UPS were abdominal pain, weight loss, fever, and night sweats. The splenomegaly was seen in 5 cases (No.1,9,10,11,19). Splenic UPS is histologically highly cellular, has marked nuclear pleomorphism and abundant mitotic activity (including atypical forms and necrosis), and has areas of spindle cell morphology.[29] Immunohistochemistry is often required to diagnose UPS. Although a specific antibody for UPS has not been identified, vimentin, CD68, α-SMA and α-antichymotrypsin were often positive in the previous studies (Table 1).[12-28] Immunohistochemistry may be useful for ruling out specific known lines of differentiation. Splenectomy is the most radical therapy. Although adjuvant radiotherapy had been well established to the UPS of soft tissue, unfortunately there was no strong evidence to splenic UPS. The principal purpose of adjuvant radiotherapy is to inactivate the microscopic

### Table 2

| T category | T criteria | N category | N criteria | M category | M criteria |
|------------|------------|------------|------------|------------|------------|
| TX         | Primary tumor cannot be assessed | N0         | No regional lymph node metastasis or unknown lymph node status | M0         | No distant metastasis |
| T1         | Organ confined | N1         | Regional lymph node metastasis | M1         | Distant metastasis |
| T2         | Tumor extension into tissue beyond organ | N2         | Regional lymph node metastasis | M2         | Distant metastasis |
| T2a        | Involves serosa or visceral peritoneum | N2         | Regional lymph node metastasis | M2         | Distant metastasis |
| T2b        | Extension beyond serosa (mesentery) | N2         | Regional lymph node metastasis | M2         | Distant metastasis |
| T3         | Involves another organ | N2         | Regional lymph node metastasis | M2         | Distant metastasis |
| T4         | Multifocal involvement | N2         | Regional lymph node metastasis | M2         | Distant metastasis |
| T4a        | Multifocal (2 sites) | N2         | Regional lymph node metastasis | M2         | Distant metastasis |
| T4b        | Multifocal (3-5 sites) | N2         | Regional lymph node metastasis | M2         | Distant metastasis |
| T4c        | Multifocal (>5 sites) | N2         | Regional lymph node metastasis | M2         | Distant metastasis |

AJCC = American Joint Committee on Cancer.
without symptoms tend to be discovered too late. Even if the tumor is discovered accidentally, judging whether the tumor is malignant or not is difficult. The specific CT characteristics of splenic UPS have not been well established. In this work, some characteristic CT findings were extracted. These were a low attenuation solid mass and a cystic component with mural calcifications adjacent to the mass, all of which were seen in 3 cases (7.0%) (Table 1). Pseudocysts that contain hemorrhage and debris make up 80% of all splenic cystic lesions, and 50% of pseudocysts have mural calcification.[11] As the histopathologic examination in this study revealed that the cystic component of this patient's splenic UPS contained necrotic tissue, this cystic lesion resembles a splenic pseudocyst. In other words, splenic UPS might accompanies pseudocysts in the result of necrosis of tumor. Calcifications on CT have also been noted in the setting of an abdominal UPS in another literature.[16] Of the 43 abdominal UPS discussed, 7 (16%) had intrasplenic calcifications on a preoperative abdominal CT, and ring like calcifications were seen in 3 cases (7.0%). The co-existence of a low attenuation solid mass and a cystic component with mural calcification may be an important hint to consider splenic UPS. Prior literature showed that UPS is FDG avid on FDG-PET/CT,[33–35] which appears to apply to splenic UPS as well.[27] CT, MRI, and FDG-PET/CT as well as fine needle biopsy might be useful. The prognostic feature and characteristic CT findings extracted in this work do not present strong evidence due to the small-size case series, which is a limitation of this work; however, we think these findings are worth the suggestion. This is the first case report of long-term recurrence-free survival following surgical treatment of a splenic UPS.

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