Laparoscopic Resection of an Undifferentiated Pleomorphic Splenic Sarcoma

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ABSTRACT

Background: Splenic tumors are rare. Malignant fibrous histiocytoma (MFH) of the spleen is one of the least common primary splenic tumors. Review of the literature shows that a laparoscopic resection has never been tried.

Method: We discuss the case of a 76-year-old man with a 7-cm MFH in the spleen and present a review of splenic sarcomas.

Results: The patient underwent a successful laparoscopic splenectomy; pathology revealed a rare undifferentiated pleomorphic sarcoma of the spleen. A review of the international literature identified 15 additional cases of primary splenic MFH. Survival was rarely longer than 15 months.

Conclusion: Malignant fibrous histiocytoma of the spleen is an exceedingly rare tumor with a poor prognosis. In experienced hands, laparoscopic splenectomy is a feasible operative choice for primary splenic sarcoma.

Key Words: Undifferentiated pleomorphic sarcoma, Malignant fibrous histiocytoma, Laparoscopic splenectomy.

INTRODUCTION

Laparoscopic splenectomy (LS) is one of the many successful applications of minimally invasive surgical techniques. Since the first laparoscopic splenectomy performed in 1991 by Delâtre, this approach has been adopted as the procedure of choice to treat benign splenic pathologies. However, data on laparoscopic splenectomy for malignant tumors is scarce because of the rarity of the occurrence of primary malignant tumors in the spleen. These tumors can be classified broadly as lymphoid and nonlymphoid. Non-Hodgkin’s lymphoma is the most common primary lymphoid tumor, and angiosarcoma is the most common nonlymphoid malignant neoplasm. The remaining nonlymphoid tumors, such as hemangioendothelioma, malignant fibrous histiocytoma (MFH), fibrosarcoma, and leiomyosarcoma, are exceedingly rare and are only anecdotally reported. This report presents the first case of splenic malignant fibrous histiocytoma treated by LS reported in the international literature.

CASE REPORT

A 76-year-old man presented to the Mayo Clinic with a 4-week history of left upper quadrant abdominal and flank pain. He had an intentional weight loss of 30 pounds over the past 18 months. A gastrointestinal review of systems was negative. He had no fever, sweats, or chills. His past medical history was significant for prostate cancer, recurrent kidney stones secondary to cystinuria, hypertension, coronary artery disease, and hyperlipidemia. His past surgical history included multiple stone extraction procedures, including bilateral open stone extractions. He underwent radical retropubic prostatectomy and pelvic lymphadenectomy in 1997.

On physical examination, he was well appearing, afebrile, and normotensive. His body mass index was 25kg/m². His abdomen was soft, nondistended, slightly tender to palpation in the left upper quadrant; no masses or organomegaly were appreciated on deep palpation. Laboratory data showed a white blood cell count of 7,300/L, hemoglobin of 10.6g/dL, and platelets of 241,000/L. His creatinine was 1.4mg/dL.
An outside CT scan of the abdomen and pelvis with oral and intravenous contrast showed a 4.8 x 6.1 x 5-cm heterogeneous splenic mass that was not present on a CT scan 10 months earlier (Figure 1). The mass had irregular borders and contained enhancing nodular and cystic components. It was reported as suspicious hemangioendothelioma, angiosarcoma, hemangiopericytoma, and other malignant processes, such as metastases or possibly infection. An incidental splenule was noted in the splenic hilum.

An ultrasound-guided biopsy of the splenic mass revealed a high-grade malignant neoplasm, unable to be further classified on cytology. A PET scan showed a centrally hypometabolic and peripherally hypermetabolic lesion in the spleen with a small central hematoma consistent with the biopsy done the day before. (Multiple foci of skeletal malignant type FDG uptake with associated subtle sclerosis were seen on L1, L3, left sacrum left iliac bone, left anterior superior iliac spine, and subtrochanteric left femur (Figure 2). There was circumferential uptake within a couple of loops of small bowel in the left lower quadrant that was considered most likely physiological. A contrast enhanced CT enterography confirmed noninvolvement of the bowel but showed splenic mass enlargement to 7.1 x 5.3 cm. Upper endoscopy and colonoscopy identified a cecal tubulovillous adenoma without evidence of malignancy. A carcinoembryonic antigen level was 0.6 ng/mL (normal), and PSA was <0.10 ng/mL.

Laparoscopic splenectomy, including the splenule was performed without any intraoperative complications. The patient had an uneventful postoperative course and was discharged on postoperative day 2. The final pathology confirmed an undifferentiated pleomorphic sarcoma, high grade (4 of 4). Postoperatively, a CT-guided biopsy of the left iliac bone lesion was negative for malignancy. The patient declined any treatment that would be based on the presumption of residual disease.

**DISCUSSION**

In 1881, Theodor Billroth performed the first splenectomy for sarcoma in a 43-year-old woman with lymphosarcoma.2 She died 6 months later from recurrent disease. Since that time, splenectomy has remained the preferred treatment for splenic sarcomas. Malignant fibrous histiocytomas are rare and have been classified into 5 subtypes: pleomorphic, inflammatory, myxoid type, giant cell, and angiomatoid.3 They are most commonly found in the extremities. Intraabdominal locations account for approximately 20% of MFH.4–6

We have found only 15 cases of splenic MFHs reported in the international literature (Table 1).7–18 Thirteen of the patients were treated with splenectomy, one patient had the tumor found on autopsy, and one patient did

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**Figure 1.** CT scan of the abdomen showing a 4.8-cm AP x 6.1-cm width x 5-cm length nonhomogeneous mass in the spleen. This mass has an irregular outline and contains several enhancing nodules and cystic spaces.

**Figure 2.** PET of the pelvis showing multiple foci of skeletal malignant-type FDG uptake, including left sacrum at the level of the S2 foramen and left iliac bone adjacent to the SI joint.
not receive any treatment. Patient ages ranged from 11 years to 82 years, with a mean age of 53. The reported survival ranged from 9 days to 18 months. Radiation was given to one of these patients after splenic resection, and chemotherapy was given to one patient. One patient was treated with combined chemotherapy and radiation. The splenic mass ranged from 375g to 1850g (mean, 1136). The tumor size ranged from 2.5cm to 21.5cm (mean, 12.9). To our knowledge, this is the first reported case of a splenic pleomorphic sarcoma treated by laparoscopic splenectomy.

Laparoscopic splenectomy has become progressively accepted as an advantageous and safe approach for splenectomy. It has been shown to have reduced morbidity and mortality rates compared with open splenectomy. 

| Study                | Case | Age | Gender | Spleen Size | Tumor Size (cm) | Subtype          | Treatment                              | Survival                  |
|----------------------|------|-----|--------|-------------|-----------------|-----------------|----------------------------------------|---------------------------|
| Hashmi (2009)        | 1    | 76  | M      | 375g        | 7.1 x 5.3       | Pleomorphic     | Laparoscopic splenectomy               | To date                   |
| Govoni (1982)        | 2    | 51  | F      | 1400g       | 21 x 25 x 10    | Pleomorphic     | Open splenectomy                       | Alive at 7 months post op|
| Wick (1982)          | 3    | 48  | M      | 1184g       | 8               | Inflammatory    | Open splenectomy plus radiation        | Alive with liver mets at 18 months   |
|                      | 4    | 51  | F      | 1400g       |                 | Inflammatory    | Open splenectomy                       | Alive at 17 months        |
|                      | 5    | 54  | M      | 1235g       |                 | Unknown         | Open splenectomy                       | Alive at 3 months         |
| Mallipudi (1998)     | 6    | 73  | W      | 15 × 10 × 8490g | 10              | Pleomorphic     | Open splenectomy                       | 19 months                 |
| Colovic (2001)       | 7    | 45  | F      | 14 × 12 × 7 | 11 × 10 × 7     | Pleomorphic     | Open splenectomy                       | 15 months                 |
| Bonilla (1994)       | 8    | 42  | F      |             |                 | Unknown         | Splenectomy, XRT, chemotx              | 4 months                  |
| Katsuura (2006)      | 9    | 82  | M      | 13cm        | 2.5 × 3         | Inflammatory    | Open splenectomy                       | Alive at 18 months        |
| Lieu (1993)          | 10   | 71  | M      | 1870g       | 9.5             | Pleomorphic     | Open splenectomy                       | 9 days                    |
| Ozaras (2003)        | 11   | 51  | F      |             | 12 × 11 × 10    | Pleomorphic     | Open splenectomy                       |                           |
| Sieber (1990)        | 12   | 41  | M      | 28×17×12 1850g | 21.5           | Pleomorphic     | Open splenectomy                       | 6 months                  |
| Yu (1989)            | 13   | 11  | F      |             | 15 × 15 × 10    | Inflammatory    | Open splenectomy                       |                           |
| Jinno (1987)         | 14   | 53  | M      |             |                 | Pleomorphic     | Autopsy                                |                           |
| Bruneton (1988)      | 15   | 54  | M      | 20 cm       |                 | Unknown         | Open (partial) splenectomy             |                           |
| Yuan (2006)          | 16   |     |        |             |                 |                 | None                                   |                           |

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tomy in many published series (morbidity 19% vs. 56%; mortality 2% vs. 18%). Most reports include patients with benign diseases and small spleens. LS for malignant disease can be a greater challenge, because of the size of the spleen and the general condition of the patient. Burch et al\textsuperscript{19} compared the outcomes of LS for benign versus malignant neoplasms. They showed that there was no statistically significant difference identified between those undergoing LS for benign versus malignant disease in terms of length of stay, complication rate, or mortality. Although there were significant differences between the 2 groups in terms of operative time, splenic weight, and the need for an accessory incision for spleen retrieval.

One criticism of the laparoscopic approach has been the lack of tactile feedback and perhaps an inability to identify accessory splenic tissue. The incidence of accessory spleen is approximately 15%.\textsuperscript{20} Failure to detect and ablate accessory splenic tissue may lead to treatment failure in case of malignancy. Studies show that the lack of the ability to palpate does not compromise the surgeon’s ability to find and remove accessory splenic tissue when LS is compared to OS.\textsuperscript{21,22}

Patients with splenic sarcoma may be at risk for operative conversion to an open operation for bleeding. In fact, hemoperitoneum due to splenic rupture is seen in up to 13% to 30% of cases and is often the first manifestation of the disease.\textsuperscript{25}

Maintaining the integrity of the capsule is advisable for both oncological and hemostatic reasons to minimize the chance of tumor dissemination or bleeding from the parenchyma. It is critical to avoid iatrogenic splenic rupture. Therefore, splenic size and surgeon experience are key determining factors when deciding on an open, a hand-assisted, or a total laparoscopic splenectomy.

**CONCLUSION**

Primary malignancies of the spleen are rare and are primarily treated with splenectomy. This case report shows that laparoscopic splenectomy is a feasible operative choice for patients with sarcomas, allowing the advantages of laparoscopic surgery and a quicker healing process permitting earlier institution of adjuvant treatment if necessary. The large size and increased risk of bleeding will likely be the greatest challenges for surgeons performing laparoscopic splenectomy for splenic sarcomas.

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