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Case report

Robot-assisted laparoscopic removal of an extraperitoneal pelvic solitary fibrous tumor

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1. Introduction

Solitary fibrous tumors (SFTs) are rare, typically benign, fibroblastic mesenchymal soft tissue tumors (Davanzo et al., 2018). They have been well-studied in the pleura, but the occurrence of these tumors in extra-pleural sites has been more appreciated over the last few years. Approximately 30% of cases have been found within the peritoneal cavity, retroperitoneal soft tissues, or pelvis (Ronchi et al., 2018).

Several case reports of pelvic SFTs have been published; however, there are limited reports of SFTs within the gynecologic literature (Yamada et al., 2019). Pelvic SFTs remain a diagnostic challenge. Because of their rarity, they are poorly recognized by clinicians (Davanzo et al., 2018). Gynecologic surgeons should be aware of pelvic SFTs despite their relatively rare incidence, as they have been reported in vulva, vagina, uterus, cervix, paravaginal space, bladder, and breast.

This case demonstrates the typical morphology, histologic features, and characteristics of SFT and highlights specific diagnostic and surgical challenges due to its unusual location within the pelvis.

2. Case presentation

This patient was a 52-year-old, post-menopausal, nulliparous woman who initially presented to a community emergency department (ED) with intermittent crampy lower abdominal pain, bloating, and nausea. At that time, she had no associated fevers, bladder or bowel complaints, weight loss, or vaginal symptoms. She had a computed tomography (CT) scan of her abdomen/pelvis that demonstrated a 3.5 × 2.8 × 4.0-cm indeterminate, complex right lower pelvic mass, which was clearly separate from the right ovary. She was referred to a community gynecologist and general surgeon. Due to concern for possible malignancy, she was referred to surgical oncology at our institution though was lost to follow-up for the next three years due to financial barriers.

She presented again for care through a community ED for continued intermittent lower abdominal pain, with new development of pelvic pressure and urinary frequency. CT scan of abdomen/pelvis at that time demonstrated interval growth of this mass, now measuring 6.1 × 5.7 × 7.0-cm. She was referred to gynecologic oncology at our institution for re-evaluation.

2.1. Diagnostic work-up

On examination, she was found to have a firm right-sided paravaginal mass palpable on bimanual exam. Her cervix appeared grossly normal, and the uterus was freely mobile. The diagnostic work-up included a repeat CT scan of her abdomen/pelvis, transvaginal ultrasound, colonoscopy, and serum tumor markers.

The CT scan of abdomen/pelvis showed an indeterminate, complex heterogenous right-sided pelvic mass with cystic and solid components (Fig. 1A-C). It had ill-defined margins and abutted the right pelvic side wall. The mass showed interval growth, measuring 6.7 × 5.0 × 7.8-cm, and remained separate from the uterus and right ovary (Fig. 1A-D). There was leftward deviation of the bladder, right ureter, and uterus due to mass effect, without evidence of invasion. She was also noted to have pelvic lymphadenopathy with some lymph nodes measuring greater than 1-cm, which were stable from prior examination.

Transvaginal ultrasound noted an unremarkable uterus measuring 6.8 × 3.8 × 4.5-cm with endometrial thickness of 8-mm (Fig. 1E) and a normal left ovary. No normal right ovarian tissue was visualized, though

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a septate complex mass with cystic and solid components measuring 7.0 × 4.9 × 7.5-cm was identified in the right adnexa (Fig. 1F).

The patient underwent colonoscopy, which was unremarkable. Tumor markers, including CA-125, CEA, and CA 19–9, were within normal limits. Given the unclear etiology of this mass, additional imaging such as magnetic resonance imaging (MRI) was considered; however, this patient experienced financial and insurance barriers prior to hospital admission. Additionally, needle biopsy of the mass and / or enlarged lymph nodes was considered; however, our team opted for a multi-disciplinary definitive surgical approach as the patient strongly desired complete resection. The final pre-operative assessment was an enlarging complex pelvic mass of uncertain origin in the paravaginal/paravesical region. She was evaluated by surgical oncology and urology teams in consultation for surgical planning.

2.2. Surgical approach and findings

Robot-assisted laparoscopic excision of this mass was planned with combined gynecologic oncology and urology teams. We believed robotic surgery in this case offered advantages of magnification and access to a space that would have been difficult to visualize with an open approach.

The patient was positioned in dorsal lithotomy. The procedure began with cystoscopy, which noted external compression from the bladder on the right side, though otherwise was unremarkable. The abdomen was entered directly with a 5-mm optical trocar at the Palmer's point within the left upper quadrant. Four robotic ports and a fifth assistant port were placed along the upper abdomen in the typical distribution for a hysterectomy for use of the camera and three arms of the robot. The patient was then placed in steep Trendelenburg.

On laparoscopy, the uterus, bilateral fallopian tubes, and ovaries were grossly normal appearing. The mass was found in the retroperitoneum deep within the right paravesical space, measuring 7–8-cm in largest diameter. The mass was noted to have cystic and solid components and was densely adherent to surrounding structures near the right external iliac vessels, right lateral bladder, and right obturator nerve. Significant neovascularity around the tumor was also noted, making for very tedious and challenging dissection. Despite these findings, no tumor invasion into surrounding structures was identified.

The pelvic mass was identified retroperitoneally after performing a pelvic lymphadenectomy. The mass was bordered laterally by the right external iliac vein and artery and bordered medially by the urinary bladder and anteriorly by the bony pelvis (Fig. 2). The ureter was noted to course laterally and posterior to the mass. Sharp and blunt dissection were used to identify the right obturator nerve inferior and lateral to the mass. The surrounding fibrous tissue and vessels were dissected away from the tumor taking care to ensure the obturator nerve was spared. The bladder was retracted medially, and the mass was noted to be firmly adherent to the right lateral bladder wall. To increase exposure, the bladder was mobilized medially by transecting the right medial umbilical ligament. This dissection was carried through into the avascular retropubic space. This was followed by circumferential dissection until the mass was completely excised (Fig. 2; Supplemental Video 1). Following tumoral resection, the bladder was reinspected cystoscopically and no injury or leak was noted. A Foley catheter was kept in place until post-operative day two for bladder decompression as recommended by our urology colleagues. The patient otherwise had an uncomplicated postoperative course and was meeting appropriate milestones for discharge by post-operative day one; however, due to difficulties with arranging transportation home, she could not be discharged until postoperative day three.

2.3. Histopathology

On gross pathology, we found a boggy semi-solid well-circumscribed mass measuring 7–8-cm in largest diameter. The mass was covered in a thin fleshy capsule and was comprised of soft, shaggy fibrous tissue with a small amount of adipose tissue and cystic components (Fig. 3A).

The mass had a unique microscopic architecture characterized by bland spindle cell proliferation with myxoid background (Ronchi et al., 2018; Tariq et al., 2021). The tumor was vascularized with delicate hyalinized vessels in a branching pattern known as "staghorn" appearance (Fig. 3B) (Davanzo et al., 2018; Gold et al., 2002; Demicco et al., 2012). Cells showed mild pleomorphism, but no significant cytologic atypia. Mitoses were inconspicuous and necrosis was not identified

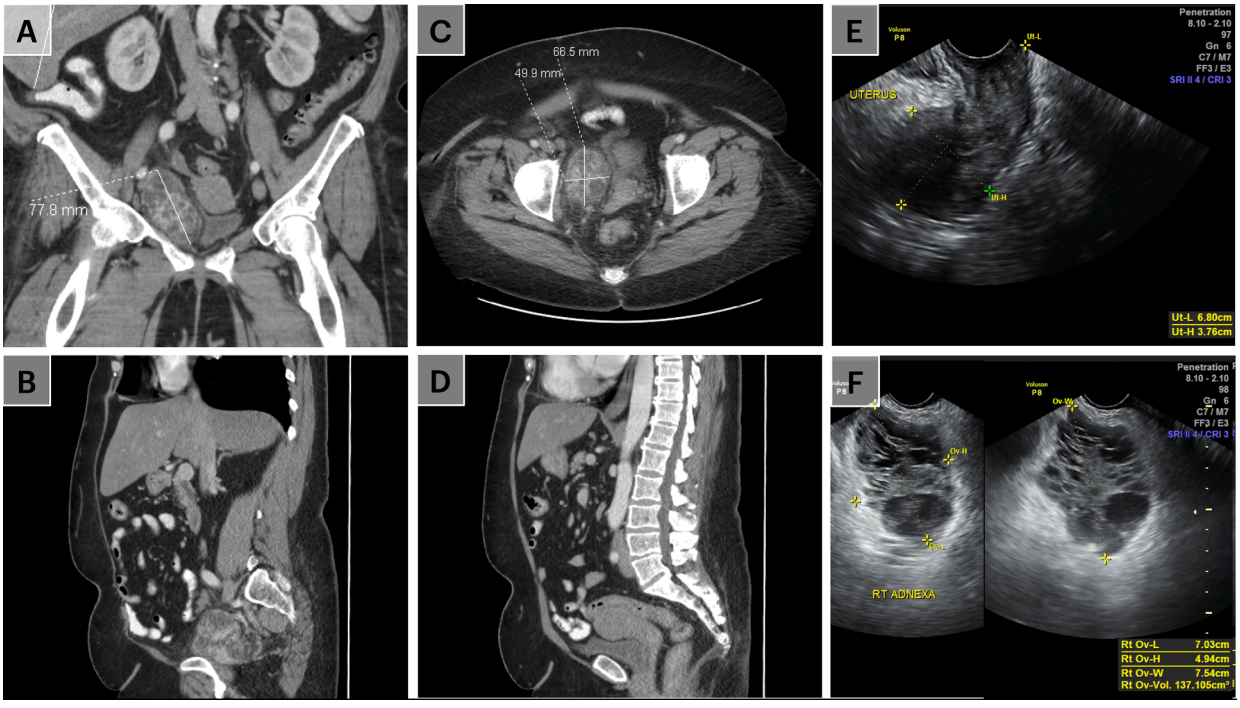

Fig. 1. Computed tomography (CT) and ultrasonography demonstrating pelvic mass. **(A)** Coronal CT of pelvic mass; **(B)** Sagittal CT view; **(C)** Axial CT view; **(D)** Sagittal CT view of normal appearing uterus; **(E)** Ultrasonography demonstrating normal appearing uterus; **(F)** Ultrasonography showing right adnexal mass.

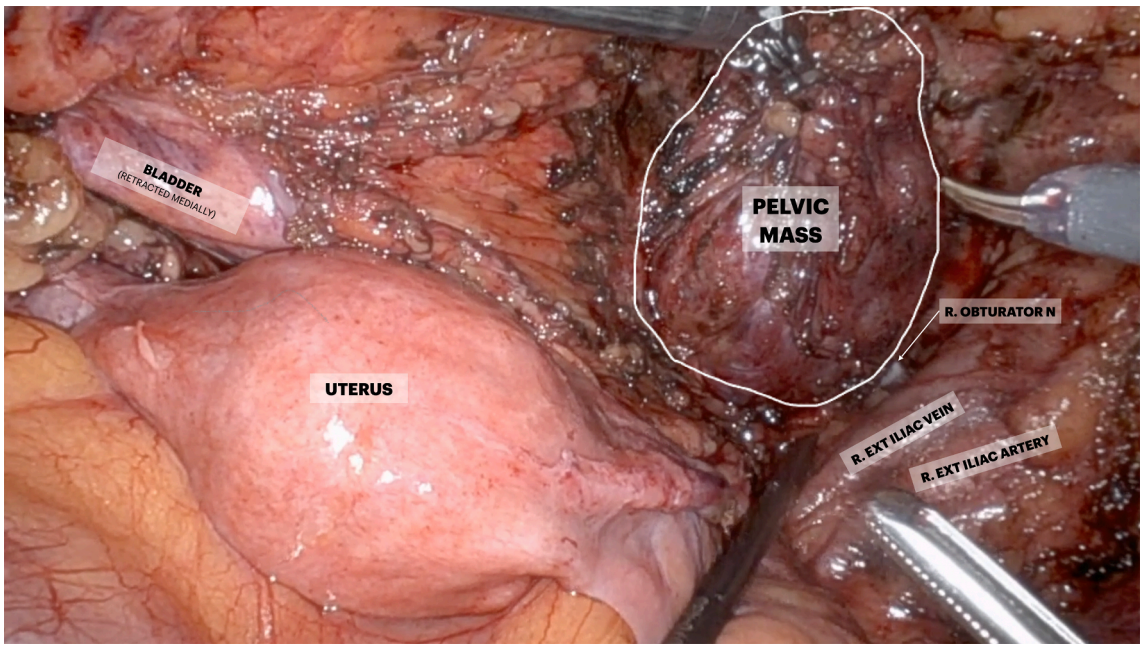

Fig. 2. View of robot-assisted laparoscopic excision of pelvic mass from right paravesical space, bordered laterally by the right external iliac vein and artery and bordered medially by the urinary bladder and anteriorly by the pelvic brim.

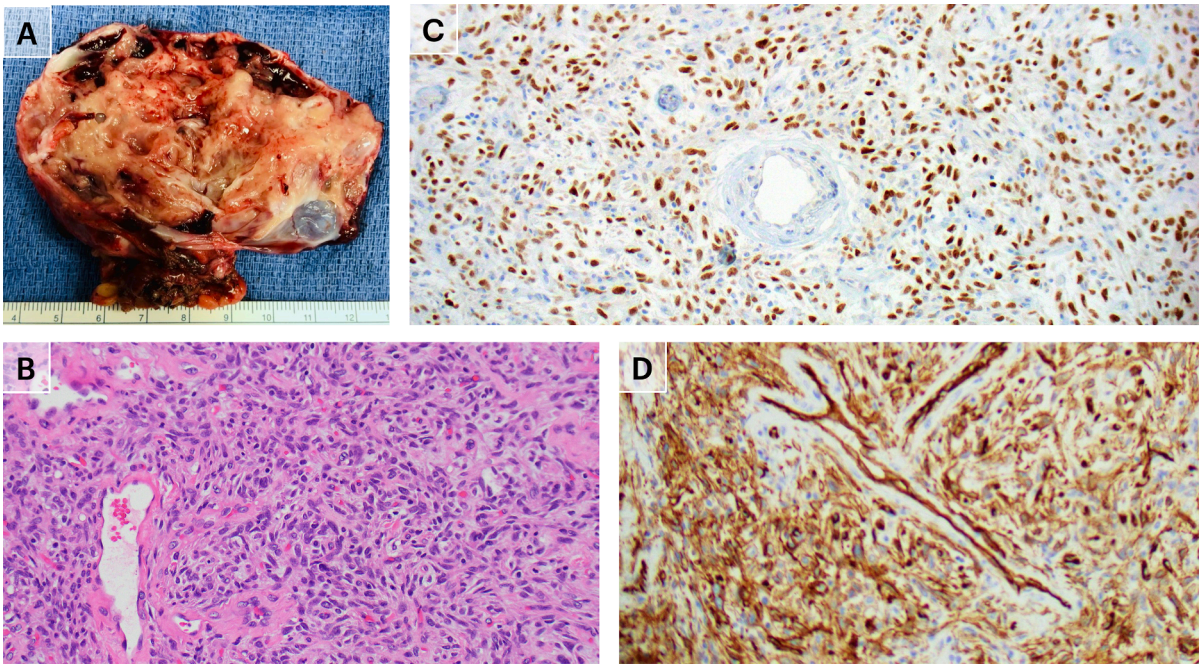

Fig. 3. Photographs of gross surgical specimen and histopathology. **(A)** View of cut surface of pelvic mass; **(B)** Photomicrograph of histological section stained with hematoxylin and eosin; **(C)** Immunohistochemical staining showing specimen stained positively for CD34 and **(D)** STAT6, consistent with diagnosis of solitary fibrous tumor.

(Fig. 3B). A focally positive margin was noted; however, this may have been artifactual. The two pelvic lymph nodes excised were negative for malignancy.

Immunohistochemistry is helpful for distinguishing SFTs from other differential diagnoses such as fibroma or leiomyoma (Doyle et al., 2014). The vascular network of this tumor is highlighted by cluster of differentiation 34 (CD34) staining (Fig. 3D) (Flint and Weiss, 1995). These tumor cells also demonstrated nuclear expression of STAT6 protein, which is a highly sensitive and specific marker for SFTs (Fig. 3C) (Doyle et al., 2014; Yoshida et al., 2014). This pattern, in conjunction with the tumor morphology, is compatible with a SFT.

3. Discussion

This patient's presentation is typical of what is currently known regarding SFTs. These tumors most commonly arise in the fifth to seventh decade of life (Tariq et al., 2021; Demicco et al., 2012). Patients often present with symptoms related to mass-effect (Davanzo et al., 2018; Daigeler et al., 2006). Pre-operative diagnosis can be very challenging to clinicians and radiologists, as it is often poorly recognized and

SFTs can mimic malignant tumors originating from gynecologic organs when they develop in the pelvic retroperitoneum (Yamada et al., 2019).

The behavior of extra-pleural solitary fibrous tumors can be unpredictable. About 10–25% of these tumors can show malignant behavior in the form of recurrence or metastatic disease. Features typically seen with malignant SFTs include large tumor size, increased mitotic index (greater than 4 mitoses per 10 high-power field), variable cytologic atypia, tumor necrosis, and infiltrative margins or incomplete surgical excision (Gold et al., 2002; Demicco et al., 2012; England et al., 1989). However, these indicators are not completely reliable as there are reports of some SFTs with a benign microscopic appearance behaving aggressively following complete surgical excision, while others with more malignant seeming histological features may not (van Houdt et al., 2013; de Perrot et al., 2002). The mass in this patient would be considered benign, or low risk, given the absence of features typically seen with malignant SFTs.

Treatment of SFTs requires complete surgical excision (Kayani et al., 2018). A robotic approach was highly effective given the difficult location of this pelvic tumor. The utility of chemotherapy and radiotherapy has been considered for incomplete resection or recurrence, though the efficacy/value of adjuvant therapy is uncertain (Davanzo et al., 2018; Park et al., 2013). Approximately 10–25% of these tumors will recur and have potential to metastasize, though the ten-year disease specific survival rate is 73–100% (Gold et al., 2002; Demicco et al., 2012; Daigeler et al., 2006).

There are no current formal recommendation guidelines for surveillance of SFTs, though continued long-term follow-up is recommended given possibility of late recurrence (Davanzo et al., 2018; Park et al., 2013). This case was reviewed at our institution's gynecologic oncology tumor board conference regarding further management. Given that the lungs are the most cited location of distant metastasis (Kayani et al., 2018), we hope to follow this patient initially with chest imaging every 3-months and CT abdomen/pelvis every 3–6 months for the next two years. Due to this patient's socioeconomic challenges returning regularly to our institution, our team plans to involve her primary care physician in her surveillance.

4. Patient consent statement

Written informed consent was obtained from the patient for publication of this case report, accompanying images, and film. A copy of the written consent is available for review by the editorial team upon request.

CRediT authorship contribution statement

Lauren L. Siewertsz van Reesema: Conceptualization, Methodology, Project administration, Writing – original draft, Writing – review & editing, Visualization. **Megan L. Hutchcraft:** Conceptualization, Methodology, Writing – review & editing, Project administration. **Nicholas A. Freidberg:** Conceptualization, Methodology, Writing – original draft, Writing – review & editing, Visualization. **John Lee Graves:** Writing – original draft, Writing – review & editing, Visualization. **Molly M. Tovar:** Data curation. **Prakash K. Pandalai:** Supervision. **John Roger Bell:** Supervision. **Charles S. Dietrich:** Conceptualization, Methodology, Writing – review & editing, Supervision.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Appendix A. Supplementary material

Supplementary data to this article can be found online at https://doi.org/10.1016/j.gore.2023.101198.

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