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Case report

# Surgical management of a large neurilemmoma-like leiomyoma of the uterine cervix mimicking a retroperitoneal tumor

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## 1. Introduction

Multiple tumors are derived from the smooth muscle of the uterus and can be classified into pathologies with benign or malignant behavior. Tumor subtypes with unusual histological patterns have also been described, including leiomyomas with schwannoma-like or neurilemmoma-like differentiation. Because these tumors are very rare, preoperative diagnosis is difficult, and their characteristics are often only determined after surgery and histological study (Gisser and Young, 1977). Specifically, these tumors exhibit a regular, spindle-shaped cellular pattern that suggests a peripheral nerve sheath tumor (Gisser and Young, 1977; Kempson and Hendrickson, 2000).

Herein, we illustrate a tumor that had not been pre-operatively diagnosed and simulated a retroperitoneal tumor. This tumor required a difficult surgical approach, and subsequent histology demonstrated that it simulated a schwannoma.

### 1.1. Presentation of case

A 46-year-old woman had defined parity and no history of exposure to toxins, chemotherapy, or radiotherapy. She consulted our hospital for abdominal distention, slight vaginal bleeding, hyporexia, quick satiety, and constipation since approximately 6 months. Abdominal ultrasonography revealed a large, solid mass with mixed echogenicity and a cystic component occupying the abdominal and pelvic cavities. The tumor measured more than 30 cm in diameter and displaced the liver and spleen; ascites was not observed. Abdominal computed tomography revealed a large tumor (31.4 cm in diameter) that displaced the intestinal loops toward the anterior and upper region of the abdomen without signs of intestinal obstruction (Fig. 1A). The tumor surrounded and displaced the pelvic body of the uterus toward the abdominal cavity; the myometrium appeared normal, and the endometrium was homogeneous and measured 6 mm. The pelvic cavity and the entire abdomen were occupied by the tumor till the level of the kidney poles (Fig. 1B). No metastatic lesions were evident in the liver, spleen, pancreas, or kidneys. The results on thoracic computed tomography were normal. Colonoscopy revealed extrinsic compression of the left colon, and the esophagogastroduodenoscopy results were normal. The levels of tumor markers were as follows: CA 125, 29 U/ml; CA 19-9, 28 U/ml; and CEA, 2.13 U/ml, all of which were within the normal ranges.

Physical examination revealed an Eastern Cooperative Oncology Group (ECOG) performance status of 1, with a large abdomino-pelvic mass that occupied the entire abdomen and reached the epigastrium. The tumor was not mobile, and adenopathy was absent. Gynecological clinical examination revealed a uterine cervix that had collapsed owing to the tumor, and the exploration of the parametrium was difficult because of the large tumor and compression of the mass toward the pelvis. Endocervical or endometrial biopsy was not performed because it was technically impossible.

The patient underwent exploratory laparotomy for possible gynecological tumor resection originating from an unknown primary organ or retroperitoneal tumor.

A vertical laparotomy was performed due to the large tumor volume. The surgical findings identified a solid, lobed, and highly vascularized tumor with a maximum diameter of 40 cm. This tumor was not mobile, and it bilaterally extended toward the infundibulo-pelvic vascular pedicles; it involved the uterus and adnexes. A uterine body without tumor was clearly differentiated, and the round ligaments and wide ligaments were elongated but were not the origin of the tumor; the ovaries and fallopian tubes appeared normal (Fig. 2A). Moreover,

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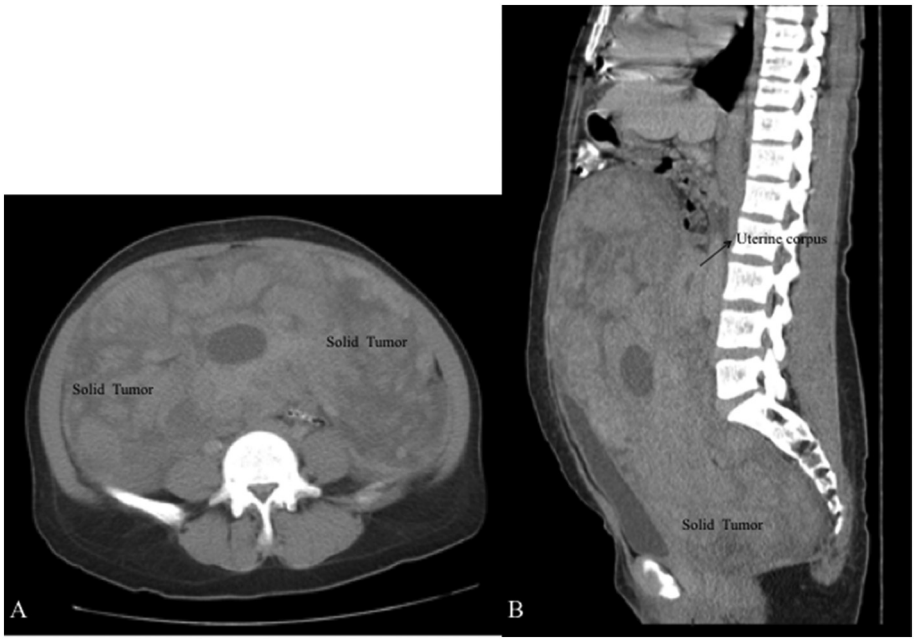


**Fig. 1.** A. Abdominal computed tomography showing a large heterogeneous abdominal tumor. B. The tumor reaching the level of the kidney poles, causing an ascended uterus and compressing the bladder.

adhesions were identified in the small intestine, left colon, lateral pelvic peritoneum, bladder, ureters, and right external iliac and hypogastric vessels.

The bladder was raised on the anterior face of the tumor, and the sigmoid colon and rectum remained in contact with the posterior portion of the tumor until the lumbosacral level but had not been infiltrated by the tumor (Fig. 2B). The ureters were not in their anatomical location, appeared dilated, and were attached to the lateral tumor faces. Tumor adhesions without infiltration were evident mainly on the external right iliac artery and adjacent lymph node tissue. The tumor did not seem to affect other organs, and suspected lymph nodes metastases in the pelvic or aortic regions were not evident. The tumor, uterus, ovaries, fallopian tubes, adhesions, and right external and internal iliac lymph nodes were resected, and the surgery margins were tumor-free, including the upper-third portion of the vagina. The patient did not require intestinal derivation, and tests were performed to confirm rectal and vesical integrity. Preoperative hemoglobin was 10.5 g/dl, 6 units of red blood cells, 6 units of plasma and 10 units of platelets were reserved. Intraoperative bleeding of 800 $cm^3$ + 800 $cm^3$ intratumoral blood volume was estimated. A total of 3000 $cm^3$ of crystalloids + 2 units of compatible red blood cells were infused. Postoperative hemoglobin was 12 g/dl, hematocrit 38%, serum creatinine 0.64 mg/dl. No additional measures were required to maintain hemodynamic stability. The patient remained hospitalized for 4 days without complications.

Histopathology revealed a multilobed tumor weighing 5425 g, measuring 40 × 32 × 10 cm with cavitated areas; the largest such area measured 4 × 2.5 cm and contained mucoid material; necrotic and myxoid areas measured 3 × 2 cm on average. The uterine cervix was fully enveloped by the tumor. The uterine corpus measured 7 × 5.5 × 3.5 cm without any endometrial lesions, and the myometrium was 2.5-cm thick. The fallopian tubes and ovaries exhibited normal morphology. Multiple cross-sections of the tumor revealed oval and spindle-shaped cellular pattern containing eosinophilic cytoplasm arranged in patterns that resemble Antoni A areas, paucicellular areas reminiscent of Antoni B areas, and myxoid areas with multiple thick-walled vessels (Fig. 3A and B). Moreover, less than 1 mitosis was observed in 50 high-magnification fields, and areas of tumor necrosis were absent. Hematoxylin-eosin staining revealed a lesion that resembled a benign peripheral neural sheath tumor (schwannoma) but did not rule

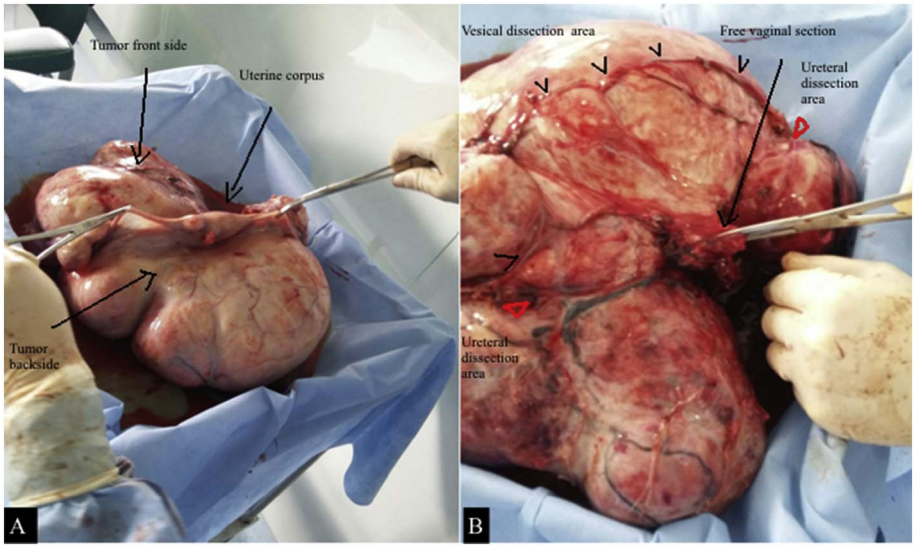


**Fig. 2.** A. A large tumor of cervical origin with a uterine body, round ligaments, and free adnexal tissue. B. Vesical, ureteral and vaginal dissection areas.

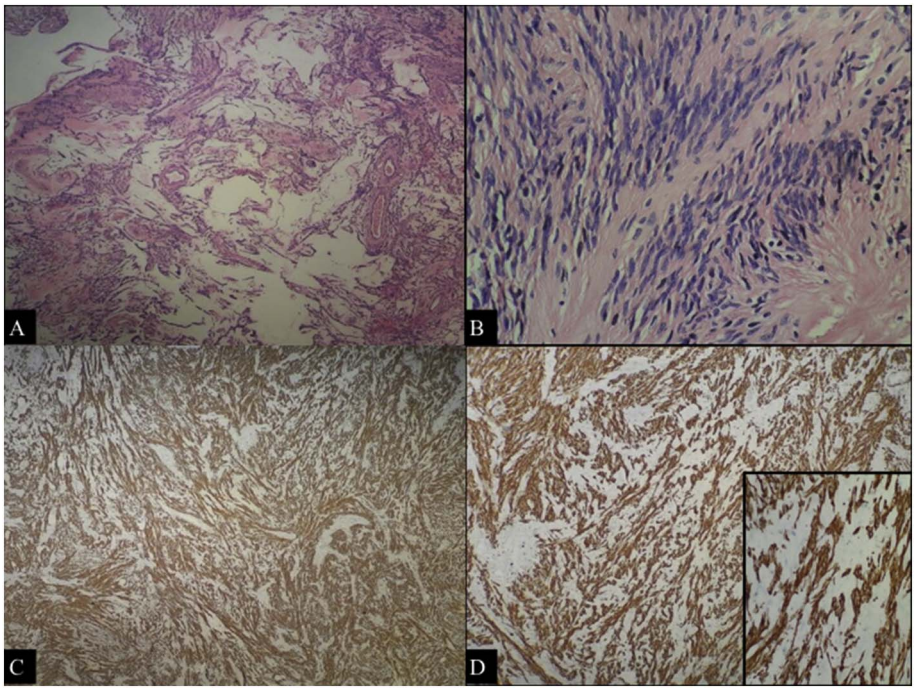


**Fig. 3.** A. Antoni A areas, paucicellular areas reminiscent of Antoni B areas, and myxoid areas with multiple thick-walled vessels. B. Oval and spindle-shaped cellular pattern with eosinophilic cytoplasm. C. Positivity for smooth muscle actin D. Positivity for desmin.

out a leiomyoma with schwannoma-like differentiation. Immunohistochemistry studies revealed positivity for desmin and smooth muscle actin and negativity for S100, CD117, and DOC1 (Fig. 3C and D), a profile that rules out a benign peripheral neural sheath tumor and favors a leiomyoma with schwannoma-like differentiation, which also affects the exocervix. The lymph nodes were negative for tumor involvement. Thus, the patient was diagnosed with a leiomyoma with schwannoma-like differentiation that affected the exocervix but not the endocervix.

## 2. Discussion

Although neurilemmoma-like or schwannoma-like leiomyomas of the uterine body are very rare, they are most commonly diagnosed in the gastrointestinal tract. Therefore, a neurilemmoma-like or schwannoma-like leiomyoma in the uterine cervix is extraordinarily rare. These tumors have usually been observed in young women ranging from 25 years of age to perimenopausal age, and their symptoms are typically associated with the location and size of the tumor; in several cases, abnormal vaginal bleeding was described as a symptom. Moreover, tumor sizes ranging from 1.5 cm to 7 cm have been described (Gisser and Young, 1977), but tumors with large volume of this type of cervical origin has not been previously described.

In case reports, computed tomography (CT) or magnetic resonance imaging of the pelvis and abdomen is commonly used for detecting lesions of the uterine body, cervical polypoid lesions, and sometimes adnexal masses. In this study, abdominal CT was not enough to clearly establish the origin of the tumor; the mass was large, displaced abdominal organs, and clearly extended to the retroperitoneum. Interestingly, retrospective evaluation of imaging study findings revealed that the uterus ascended into the abdominal cavity owing to the large tumor volume, and this appearance was confirmed by the surgical findings. We believe that the surgeon may consider preoperative embolization as a reasonable option in terms of intraoperative bleeding control. Retrospectively, in the presence of a large solid abdominopelvic tumor that ascends the uterus from the pelvis and impairs cervical exploration, embolization of uterine arteries or hypogastric arteries should be considered as a pre-surgical option (Conforti et al., 2015).

The preoperative differential diagnosis included a sarcomatous tumor of uterine origin or a retroperitoneal liposarcoma, especially based on the imaging findings.

On histopathology, these tumors simulate a peripheral tumor with a neural sheath origin, with Verocay bodies and Antoni A palisade pattern or Antoni B pattern, and abundant myxoid areas with little or no mitosis (Gisser and Young, 1977; Kempson and Hendrickson, 2000). The cells exhibited a spindle-shaped pattern with nuclei, forming slightly irregular palisades, but maintaining the fine features of smooth muscle differentiation, with myofilament groups, dense bodies, and a basal layer (Kempson and Hendrickson, 2000; Mazur and Kraus, 1980). In our case, the tumor contained areas exhibiting Antoni A and other areas exhibiting Antoni B patterns with abundant myxoid tissue. Immunostaining aided the differential diagnosis of the tumor because schwannomas are often positive for antigen S100 and neuron-specific enolase (Bernstein et al., 1999; Tahmasbi et al., 2012), markers that were negative on immunostaining in our case. Furthermore, uterine cervix schwannomas have been associated with neurofibromatosis type 1 (NF-1) and polypoid tumors (Keel et al., 1998), clinical features that were not present in this patient. Schwannomas (neurilemmomas) are tumors that arise from the nerve sheaths and the retroperitoneal localization is unusual with less than 5% in frequency. They have a good prognosis if complete surgical resection is achieved, otherwise they tend to relapse (Borghese et al., 2002). Cases of malignant schwannomas of the cervix report disease-free survival of up to 10 years with hysterectomy (Lallas et al., 1999). On the other hand, cervical leiomyomas are very uncommon and are estimated to represent 0.6% in hysterectomy specimens. Neurilemmoma like leiomyoma is a very rare benign tumor with a good prognosis and represents a histological variant of a leiomyoma, therefore, complete surgical resection with free tumor margins is considered the definitive treatment (Tiltman, 1998). So we did not consider giving adjuvant management of chemotherapy or radiotherapy in our case. Finally, a case of a leiomyosarcoma originating from a pre-existing neurilemmoma-like leiomyoma has been described (Kir et al., 2003); because this tumor implies a worse prognosis, the entire large resected tumor was fully processed and evaluated to corroborate the diagnosis and disregard associated dedifferentiation.

Currently, the patient has completed 15 months of follow-up and has not exhibited disease recurrence.

## 3. Conclusion

A rare large-volume tumor originating in the uterine cervix simulated a retroperitoneal tumor. Cervical tumors with rare histologies remain a surgical challenge for surgeons and a diagnostic challenge for pathologists.

### 3.1. Consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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## Conflict of interest

There are no conflicts of interest.

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