Intravenous Endometrial Stromal Sarcoma: a case report

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Endometrial stromal sarcoma (ESS) is a rare mesenchymal neoplasm that usually occurs as a primary tumor of the uterus. Endometrial stromal sarcomas may occasionally arise in other sites, such as the ovary, mesentery, and vagina. We report an endometrial stromal sarcoma arising from the left ovarian vein after a hysterec- tomy 29 years earlier because of uterine myomas. To the best of our knowledge, it is extremely rare for endometrial stromal sarcoma to grow in the great vessels, especially the left ovarian vein. Even though this tumor is rare, it should be included in the differential diagnosis when a longitudinal mass is seen in the left lower abdomen of a female patient. Therefore, the correct preoperative diag- nosis can be established.

Endometrial stromal sarcoma (ESS) is a rare malignant mesenchymal tumor that usually develops in the uterus and occasionally arises at various extra-uterine sites [1, 2]. The overall five-year survival rate for ESS exceeds eighty percent. Approximately fifty percent of the patients, however, have tumor recurrence, mostly after a long latency period. Our case is a female patient who has undergone a hysterectomy 29 years earlier and an extrauterine ESS in the left ovarian vein is found. It is difficult to determine whether the tumor is a primary extrauterine ESS or a late recurrence of ESS. We report this case and review of the literature.

CASE REPORT

A 78 year-old female suffered from dull abdominal pain (without relationship with meals) and mild abdominal distention in recent weeks. She had undergone a hysterectomy 29 years earlier for uterine myomas. She was multipara and had no history of endometriosis or taking hormonal drugs. Physical examination, routine laboratory tests, and chest and abdominal roentgen films were within normal limits. Abdominal sonography showed a longitudinally hypoechoic mass in the left lower abdomen (Fig. 1). The further abdominal CT scanning revealed a lobulated mass with heterogeneous enhancement among the bowel loops in the left side abdomen (Fig. 2, 3a). Surgical intervention was arranged due to persistent abdominal pain. At laparotomy, a left lower retroperitoneal mass was found. The mass was located close to but not truly contiguous with the small bowel and ureter. The pathologic examination showed a well-demarcated retroperitoneal tumor measuring 8.6 × 5 × 2.8cm in size with blunt bosselated outer surface and capsule-liked boundary (Fig. 4). It was loosely attached to a fallopian tube at one pole, but not to the ovary. Microscopically, the tumor showed entirely intra- venous worm-like nodular growth adhered to the vascular walls of the venous plexus, probably the left ovarian vein (Fig. 5a). It was composed of oval
to spindle cells, frequently looking as endometrial stromal cells with oval nuclei and modest to scanty cytoplasm (Fig. 5b). Sex-cord-like structures were present in some places. The mitoses in some areas were more than 4/10 HPF. No endometriosis was morphologically identified in the lesion or in the adjacent structures. Immunohistochemical studies showed that tumor cells were positive for CD10, estrogen receptor, and progesterone receptor, but negative for actin, CD34, and alpha-inhibin, leading to the diagnosis of endometrial stromal sarcoma.

Figure 1. The abdominal sonogram shows a longitudinally hypoechoic mass (arrow) in the left lower abdomen.

Figure 2. The contrast-enhanced abdominal CT shows a lobulated mass (arrow) with heterogeneous enhancement in the left lower abdomen.

Figure 3. a. The multi-planar reformation (MPR) image demonstrates that the lower portion of this tumor (arrow) abuts the adjacent bowel wall. b. The upper portion of this tumor communicates with the left renal vein (arrow) and the vessel harboring tumor mass with worm-like nodular growth corresponds to the left ovarian vein.
DISCUSSION

ESS is a rare uterine mesenchymal tumor comprising 0.2% of uterine malignancy and is the least common (<10%) type of uterine sarcomas [1]. ESSs usually develop as primary uterine tumor, but ESSs of extrauterine origins, such as the ovary, fallopian tube, pelvic cavity, abdominal cavity, omentum, retroperitoneum, vagina, vulva, and sigmoid colon, have been reported [2-6]. It is very rare that an ESS arises from the patient in her eighth decade who had undergone a hysterectomy for uterine myomas. As far as we know, only a few cases in the literature of post-hysterectomy extrauterine endometrial stromal sarcoma have been reported [7, 8]. The pathogenesis of the extrauterine ESS is not very clear. There are two hypotheses about this neoplasm. The first one is that extrauterine ESS is associated the preexisting endometriosis [2-5]. The second is the carcinogenic effect of post radiation therapy. The influence of endometrial sarcoma following radiation treatment has been described [9, 10]. It may be a de novo tumor, derived from submesothelial pluripotential mullerian cells widely distributed in the abdominal and pelvic cavities [11].

Histologically, the ESSs are subdivided into low-grade and high-grade tumors by Norris and Taylor [12, 13]. The high-grade tumor shows a lack of plexiform vasculature, frequent atypical mitotic figures with more than 10/10 HPF, and marked cytologic atypia. The low-grade tumor shows slowly growing tumors with plexiform vasculature, rare mitotic figures, and mild cytologic atypia [13]. Moreover, high-grade ESSs show destructive myometrial invasion rather than the lymphatic or vascular permeation, which occurs in a lower-grade ESS [14]. As a result, the classification of high-grade ESS is changed to undifferentiated endometrial sarcoma and the term of ESS is used for cases of low-grade stromal sarcomas only.

The ESSs typically have an indolent growth with a tendency for late recurrence with the interval of recurrence varying from 3 months to 23 years [15]. The tendency of the great vessel invasion and formation of a tumor thrombus are in inferior vena

Figure 4. The well-defined bosselated tumor is bounded by a capsule-like structure which is actually venous wall. The left retracted cord of tumor (arrow) is in the proximal venous lumen and the right forceps clamps the venous wall.

Figure 5. a. The intravenous expanding nodular growth is revealed (H&E stain). b. Typical histological pattern of endometrial stromal sarcoma is found in places of this tumor (H&E stain).
cava also well described [16, 17].

The sonographic findings of ESS are nonspecific, such as polyoid endometrial masses, endomyometrial thickening, and adnexal masses [18]. The most common CT feature of uterine sarcoma is low-density mass with necrosis [19] without specific tomographic pattern to aid in the differential diagnosis. CT is mostly used to detect distant metastases and to stage the disease. The ESS presents isointense signals relative to the myometrium on T1-weighted imaging and hypersignal on T2-weighted imaging. Moreover, it shows heterogeneous but prominent contrast enhancement because of its rich vascularity. The lesions in the form of large masses with irregular margins, multiple nodular mass formations and intramyometrial worm-like nodular extensions are frequently seen [20].

In our patient, she has undergone a hysterectomy 29 years prior to the diagnosis of ESS. Neither slides of her uterine tumor (“leiomyoma”) nor specific imaging findings of ESS can be available for review. It is hard to determine whether the mass is a new tumor or a recurrent tumor. However, neither evidence of endometriosis nor radiation therapy of this patient is noted and the tendency of late recurrence and vascular invasion of ESS has been reported. We suppose that this tumor is a late recurrent ESS. Nevertheless, the possibility of primary extrauterine ESS can’t be completely excluded.

In our case, the vessel harboring ESS with worm-like nodular growth corresponds to the left ovarian vein (Fig. 3b). The ovarian veins form a plexus near the ovary within the broad ligament and communicate with the uterine plexus. The right ovarian vein terminates in the inferior vena cava at an acute angle and the left ovarian vein terminates in the left renal vein at a right angle. Due to the communication with the left ovarian vein, left renal vein and the inferior vena cava, tumor in the left ovarian vein may extend into the IVC. Aggressive surgical treatment is recommended to prevent sudden death due to pulmonary embolism [16, 17]. In our case, the upper margin of the tumor is not free. If the correct preoperative diagnosis is established, complete tumor thrombectomy can be accomplished to prevent pulmonary embolism and to achieve a good prognosis.

**CONCLUSION**

We have reported an unusual ESS in the left ovarian vein after hysterectomy 29 years earlier and it is more in favor of a late recurrent tumor rather than a primary extrauterine ESS. Even though this tumor is rare, it should be kept in mind in the differential diagnosis when a longitudinal mass in the left lower abdomen of a female patient with a hysterectomy. Establishing the correct preoperative diagnosis and performing the tumor resection is essential to achieve a good outcome.

**REFERENCES**


靜脈長出的子宮內膜基質肉瘤：病例報告

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子宮內膜基質肉瘤（endometrial stromal sarcoma）是種少見的間質腫瘤，通常存在於子宮，但是有時也會在其他地方出現，像是卵巢、腸系膜、以及陰道。我們報導一個少見的子宮內膜基質肉瘤，長在左邊卵巢靜脈，發生在一個因為子宮肌瘤，開完子宮切除術 29 年的婦人身上。如果能在手術前就診斷出來，就能開刀切除，避免肺栓塞，使病人有更好的療後。