EJGO European Journal of Gynaecological Oncology

Schwannomas of female genitalia from a gynaecologist's perspective: report of two cases and review of the literature

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Summary

Background: Pelvic schwannomas are extremely rare. However, when located in the pelvic cavity, schwannomas are often encountered by a gynaecologist, not a general surgeon, and are misdiagnosed as gynaecologic masses. *Case Report:* Here, the authors present two cases of pelvic schwannomas that were preoperatively misdiagnosed as broad ligament fibroid. One schwannoma occurred completely in the left broad ligament and was resected by laparoscopy without any complications. The other lesion was located in the retroperitoneum and had densely adhered to the surrounding tissues; this lesion was excised by laparotomy with considerable blood loss. *Conclusions:* Schwannomas of female genitalia are very scarce and difficulty to diagnose preoperatively. Literature review revealed 63 schwannomas arising from the female genital tract in total, 73.02% (46 cases) were located in the lower genital tract, and 26.98% (17 cases) were located in upper genital tract. The treatment modality is unique depending on the location of the tumor. Complete excision is benefical for diagnosis and treatment. The procedure can be performed safely under laparoscopy.

Key words: Neurilemmoma; Laparoscopy; Pelvic neoplasms; Genital neoplasms; Female.

Introduction

Schwannomas, also known as neurilemmomas, are benign nerve sheath tumors originating from Schwann cells. These lesions were first clearly described in 1932 by P. Masson after investigations of experimental and spontaneous schwannomas [1, 2]. Most schwannomas arise from the acoustic nerve sheath. Schwannomas located in pelvic cavity are very rare and are often misdiagnosed as uterus myomas and other gynaecologic masses. The authors present two cases of schwannomas in the pelvic cavity misdiagnosed as broad ligament fibroids. They also reviewed the literature on schwannomas in female genital organs, which are often encountered by gynaecologists, not general surgeons.

Case Report

Case one

A 29-year-old female with a pelvic mass was admitted without any uncomfortable complaints. A solid mass 2.0 cm in diameter was incidentally identified by a B ultrasound in the first trimester about two years prior. The mass was not found during the uneventful term transverse lower segment cesarean operation in May 2011. The patient follow-up during the past two years revealed that the mass grew very slowly. Computed tomography scans revealed a 4.1×3.1 cm solid mass in the left side of the uterus on admission.

Laparoscopic myomectomy was arranged on February 17, 2013. A free mass approximately 4×4×3 cm completely located in the left broad ligament was noted. The mass was not connected to the uterus and had not adhered to the surrounding tissues. Six units of vasopressin were injected into the mass before surgery. The left ureter was first separated and recognized for prudence. The left broad ligament was opened by scissors. The mass was easily separated and en bloc excised. The mass appeared yellow and xanthomatous and was very soft and friable; it appeared degenerative and was removed by morcellating. The incision was sutured with 1–0 Ethicon under laparoscopy (Figure 1). The laparoscopic surgery lasted 70 minutes with approximately 30 ml blood loss. The operation procedure was recorded and is available from the first author. The intraoperative cryosection examination suggested that the lesion was a leiomyoma. However, hematoxylin and eosin stain and immunohistochemical staining revealed that the mass was a neurilemmoma: S-100 positive (+++); KI-67 positive (+), CD117 positive (+), CD34 positive (+), PR positive (+), ER positive (+), SMA negative (-), NSE negative (-), and desmin negative (-) (Figure 2).

Case two

A 63-year-old female was admitted with a 7.2×6.1 cm mass in the pelvic cavity identified by routine pelvic examination that was confirmed by a B ultrasound. The patient had no any uncomfortable complaints. The patient underwent left adnexectomy ten years prior, and the mass was not found during that surgery.

Total abdominal hysterectomy with right adnexectomy was arranged on May 2, 2013, given the mass size, surgery history and menopausal periods of 14 years. A mass seven cm in diameter protruded into the retroperitoneum near the beginning of left broad ligament and had densely adhered to the surrounding tissues. Total abdominal hysterectomy with right adnexectomy and retroperitoneal mass excision were performed with difficulty. The mass was grey with partial areas of yellow, xanthomatous ap-

Revised manuscript accepted for publication February 3, 2015

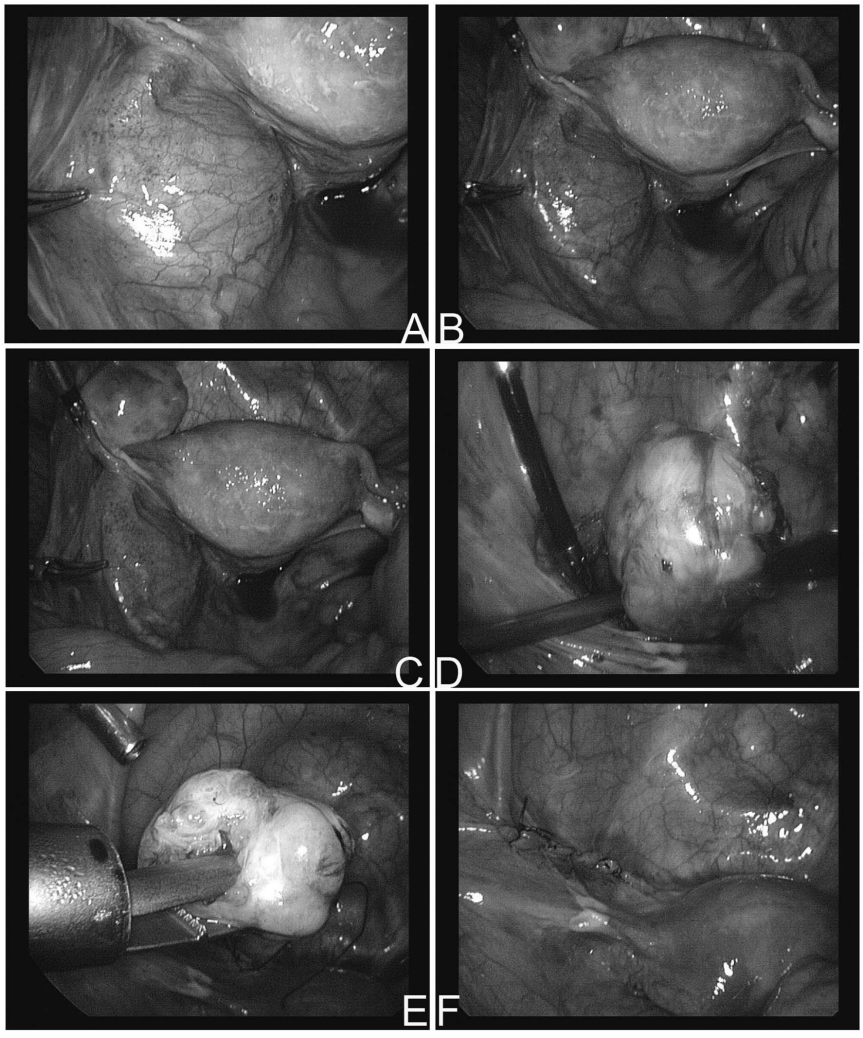

Figure 1. — Laparoscopic surgery findings of a schwannoma located in the left broad ligament. A, B, and C: location of the schwannoma mass. D and E: macroscopic appearance of the resected schwannoma mass. F: suture under laparoscopy.

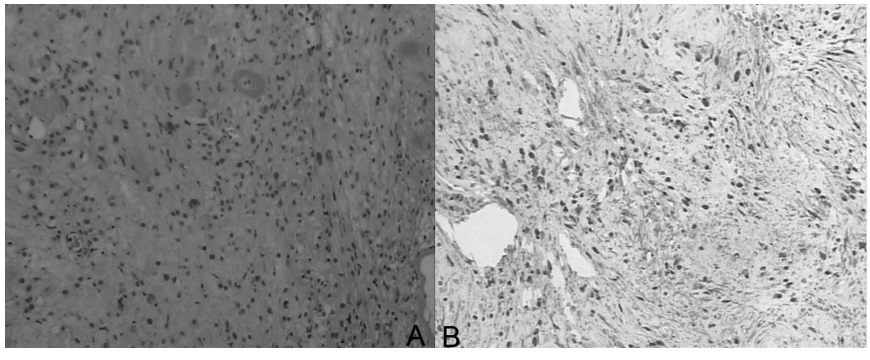

Figure 2. — A: hematoxylin and eosin-stained section of the mass shown spindle cell proliferation and thick-wall hyalinized blood vessels. (10×10 magnification). B: immunohistochemical stain shown the spindle cells strongly positive for S100 protein. (10×10 magnification).

pearance. The mass was very soft and friable and appeared degenerative. The mass had densely adhered to the presacral neurovascular plexus, and approximately 4,300 ml of blood was lost during the procedure. Blood pressure declined to 70/50 mmHg from 120/80 mmHg during the operation. The left internal iliac artery was ligated, and 12 units of leukocyte-poor red cells, 600 ml plasma, and 40 g albumin were transfused. The mass was totally excised. The surgery lasted three hours and 35 minutes. The intraoperative cryosection examination suggested that the lesion was a mesenchymal tumor. However, permanent section examination revealed not only a myoma in the uterus but also a schwannoma in the retroperitoneum: S-100 positive (+), vimentin positive (+), SMA negative (-), desmin negative (-), GFAP negative (-), CD56 negative (-), CD117 negative (-), CD34 negative (-), CD99 negative (-), CK(pan) negative (-), EMA negative (-), and HMB45 negative (-).

Table 1. — *Site distribution of schwannomas from the female genital tract.*

Site	Cases (%)	Age mean and range	Characteristics
Vulva	17 (26.98%)	34.3±17.66* 5~65	The most common presenting clinical sign was vulvar swelling. Three cases were plexiform schwannomas [8-10], one was a malignant schwannoma [11], and one was misdiagnosed as a Bartholin's gland abscess [12]. The largest size was 15×12 cm [13]. The first reported case was in 1955 [14].
Cervix	15 (23.81%)	47.92 ± 26.80* 27~73	The most common presenting clinical symptom was abnormal vaginal bleeding. Seven were malignant schwannomas [15]. The first reported case in English was in 1980 [16], but it was reported as early as in 1960 in German [17].
Clitoris	8 (12.70%)	13.5# 0~41	The most common presenting clinical sign was an enlarging clitoris. Three were plexiform schwannomas [18-20] and one was found at birth [19]. One was a malignant schwannoma, and the patient underwent radical clitorectomy and chemotherapy [21].
Vagina	6 (9.52%)	46.3# 31~62	The first vaginal schwannoma was reported in 1992 [22]. One retroperitoneal schwannoma was located at the right vaginal wall [23].
Broad ligament	5 (7.93%)	46# 42~50	The first schwannoma in the broad ligament was reported in 1959 and misdiagnosed as an ovarian cyst [24]. One was resected by laparoscopy [25].
Ovary	3 (4.76%)	71,71, and 78	The three cases were all malignant schwannomas [26-28].
Rectovaginal septum	3 (4.76%)	39, and the other two unknown	One was resected via a vaginal approach [29], the other two was via laparotomy [30, 31].
Paracolpium	2 (3.17%)	22 and 43	One was resected using the transvaginal approach [32] and the other used a suprapubic retroperitoneal operative approach [33].
Parametrium	1 (1.58%)	58	The patient underwent laparotomy with total abdominal hysterectomy and en-bloc tumor excision [34].
Fallopian tube	1 (1.58%)	40	Macroscopically, the tumor was encapsulated and pink-red with a 26×18×8-cm diameter arising as an eccentric growth from the wall of the tuba uterine [35].
Round ligament	1 (1.58%)	unknown	A giant-cell schwannoma of the round ligament [36]
Urethrovaginal septum	1 (1.58%)	unknown	A neurinoma of the urethrovaginal septum [37].
Total	63 (100%)	41.95 ± 38.43* 0~78	73.02% (46 cases) were located in the lower genital tract and 26.98% (17 cases) in upper genital tract.

* mean ± 2SD; #mean age.

Discussion

Schwannoma is a slow-growing and extremely homogeneous tumor that exclusively consists of Schwann cells. The tumor cells are always found on the outside of the nerve, but the tumor itself may either push the nerve aside and/or up against a bony structure, thereby potentially causing a few non-specific symptoms, such as dull pain, paresthesia, and motor impairment in the involved nerve. The clinical manifestations depend on the type and location of the neuronal tissue involved and the tumor size. In these two cases of neurilemmomas located in pelvic cavity, a large space was available to the tumor, allowing it to slowly grow to a large volume. In addition, no important organs occupy this area, so the tumor did not cause specific symptoms.

A definitive diagnosis can only be achieved by histopathologic examination and immunohistochemistry. S-100 protein immunohistochemistry is the most widely used staining method. CT-guided fine needle aspiration biopsy often fails to provide an accurate diagnosis, especially in condition of degenerative schwannoma [3]. Imaging modalities are unable to establish a definitive diagnosis [4]; however, these techniques can identify the mass location and its relationship to surrounding structures, thereby assisting in the selection of a suitable treatment, such as laparotomy, laparoscopy or other surgical techniques. For Case one, the size and location of the mass allowed laparoscopic surgery to be performed successfully. However, for Case two, the mass was very large and located in left side of the annex area; the patient also had a previous laparotomy history of left adnexectomy. Therefore, it was preoperatively hypothesized that the mass had adhered to the surrounding structures, so laparotomy was arranged for this patient. A considerable amount of blood was lost during surgery. Partial resection appeared to be more suitable for this patient to prevent iatrogenic damages, such as neural deficit and vessel lesions; this type of damage occurs in 10% of all cases [5]. Conservative therapy and follow-up may serve as an alternative treatment for this patient [6, 7].

Schwannomas rarely affect the female genital tract. In total, 63 schwannomas arising from the female genital tract have been reported in PubMed. Of these cases, 73.02% (46 cases) were located in the lower genital tract, and 26.98% (17 cases) were located in upper genital tract. The most affected organ was vulva (26.98%, 17/63) followed by the cervix (23.81%, 15/63). The youngest patient was identified at birth with an enlarged clitoris (Table 1).

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