

# The Pelvic Uterus-like Mass—A Primary or Secondary Müllerian System Anomaly?

Yuval Kaufman, MD\*, and Alan Lam, MD

From the Centre for Advanced Reproductive Endosurgery (Drs. Kaufman and Lam), Sydney, Australia, and The Lady Davies Carmel Medical Center affiliated to the Technion Institute of Technology–ITT (Dr. Kaufman), Haifa, Israel.

**ABSTRACT:** The pelvic uterus-like mass is a rare phenomenon in which an extrauterine mass, comprised of smooth muscle and a central cavity lined by endometrium, is found within the pelvis. The mass is associated with endometriosis and in some of the cases with congenital Müllerian malformations. There is an ongoing debate whether the finding is a result of smooth muscle metaplasia or a remnant of a Müllerian system defect. We present 2 distinct cases of a uterus-like mass. *Journal of Minimally Invasive Gynecology* (2008) 15, 494–497 © 2008 AAGL. All rights reserved.

**Keywords:** Uterus-like mass; Endometriosis; Müllerian anomaly; Smooth muscle metaplasia; Laparoscopy

The pelvic uterus-like mass is an extremely rare phenomenon in which an extrauterine mass, comprised of smooth muscle and a central cavity lined by endometrium, is found within the pelvis. The few existing reports in the literature have usually described the mass at the location of the ovary or broad ligament, often in patients with endometriosis [1–7]. In some of the cases the mass was associated with congenital malformations of the internal genitalia and the urinary system [1,2,7]. In other cases it was an isolated finding [3,5,6]. The “uterus-like mass” has been found in both extremes of the menstrual spectrum—a child near menarche [7] and a postmenopausal woman after total abdominal hysterectomy and bilateral salpingo-oophorectomy [3]. It has also been described with cases of clear cell and endometrioid ovarian carcinoma [4,8], which are the more common epithelial ovarian carcinomas associated with endometriosis. There is an ongoing debate whether the finding, termed “adenomyoma” [3], “endomyometriosis” [9], or “ovarian leiomyoma” [10], is a result of smooth muscle metaplasia (SMM) or a remnant of a Müllerian system defect. We present 2 distinct cases of a uterus-like mass.

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Corresponding author: Yuval Kaufman, MD, The Lady Davis Carmel Medical Center affiliated to the Technion Institute, of Technion Medical School, Haifa, Israel.

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## Case 1

A 39-year-old nulliparous woman presented with regular periods associated with severe dysmenorrhea, menorrhagia, mid-cycle, and acyclical pain, mainly in the right iliac fossa but at times in the left iliac fossa. The patient also frequently experienced dyspareunia and menstrual constipation. She had previously been admitted to the hospital and treated for presumed pelvic inflammatory disease. She had failed to conceive in the past but was not interested in investigating her fertility problem.

On physical examination, she was found to have a very large pelvic mass suggestive of myoma and a probable right adnexal mass. Transvaginal ultrasound scanning showed a bicornuate uterus with an enlarged left horn containing several myomas and a small right horn distended with blood and bilateral endometriomas. Computed tomography showed a bicornuate uterus with a larger left horn, absent right kidney, and a left hypertrophied kidney with normal excretion of the contrast agent.

Laparoscopy revealed a large pelvic complex adherent to the right pelvic side wall that included hematosalpinx, an endometrioma, and a suspected right rudimentary uterine horn (Fig. 1). The horn was adjacent, but not connected, to a normal sized left uterine horn. The left adnexa were normal. Pigmented and vesicular endometriotic nodules were found in the pouch of Douglas, the uterosacral ligaments, and anterior abdominal wall. Cystoscopy showed an absent right ureteric orifice. During laparoscopy the right pelvic sidewall and retroperitoneum were dissected. The

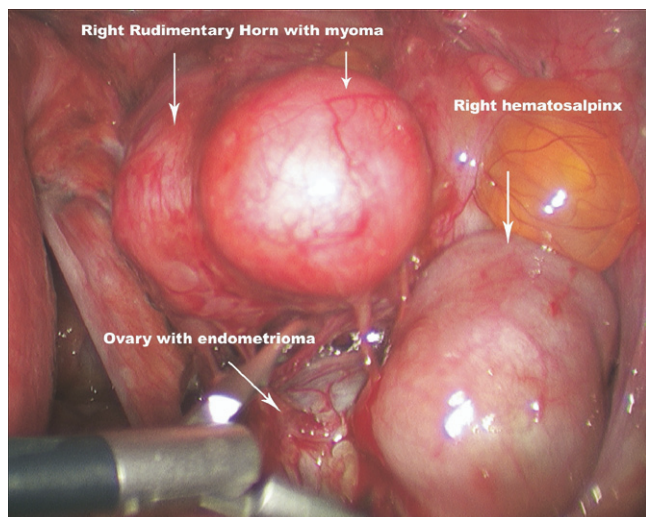


Fig. 1. In case 1, laparoscopy shows right tubal hematosalpinx, an endometrioma, and a suspected right rudimentary uterine horn.

right infundibulopelvic ligament and pelvic sidewall vessels were identified, and an absent right ureter was confirmed. The right uterine horn, ovary, and tubal mass were removed, and the endometriotic lesions were excised.

Histopathologic results showed a right 4- × 5- × 7-cm uterine horn with a central cavity surrounded by endometrium and myometrium and a 2-cm serosal leiomyoma. The cavity had no proximal tract toward the uterus. The serosal layer of the uterus was covered with fibrotic tissue. Microscopically the right uterine horn was diagnosed as adenomyosis.

## Case 2

A 57-year-old nulliparous woman was referred by her gynecologist to our clinic for management of her 3-year history of progressively worsening right iliac fossa, suprapubic and lower back pain, radiating to the right leg. The referring gynecologist had performed a right salpingo-oophorectomy when the patient was 26 and a total abdominal hysterectomy and left salpingo-oophorectomy when the patient was 38 because of endometriosis. Histopathologic results showed a normal uterus and a left tuboovarian mass containing several endometriomas and fibrovascular adhesions. The patient remained symptom free on unopposed estradiol implants until the age of 54.

On examination, she was found to have a tender mass fixed to the right pelvic sidewall. Ultrasound scanning showed an encapsulated complex mass with internal vascularity measuring 4 cm in diameter in the right iliac fossa. Her CA-125 level was normal.

Diagnostic laparoscopy showed a retroperitoneal mass firmly fixed to the right pelvic sidewall, obscuring the right ureter. An unsuccessful attempt was made to stent the right ureter. Intravenous pyelography later showed no obstruc-

tion. Computed tomography scanning showed a 5- × 5-cm mass adjacent to the right external iliac vessels. A presumptive diagnosis of endometrioma was made. The patient was referred to a vascular surgeon and a urologist for assessment. The mass was considered too risky for surgical removal. The patient was conservatively treated with oral medroxyprogesterone. After the symptoms improved and the mass remained unchanged in size for 2 years, her symptoms eventually recurred, with repeat computed tomography scanning showing the mass size doubling to 8.3 × 6.3 cm.

After careful counseling, the patient consented to definitive surgery performed in conjunction with a urologist. At laparoscopy, the retroperitoneal mass was embedded in the right pelvic sidewall, partially covered by the cecum above and firmly adherent to the bladder below. The mass appeared to be in the location of the previous ovary connected to the infundibulopelvic ligament (Fig. 2). The mass was successfully removed. Cystoscopy with stenting of the right ureter was performed at the end of the procedure. Histopathologic examination showed a 4.5- × 9- × 10.5-cm right adnexal mass consisting of smooth muscle resembling myometrium and foci of endometriosis.

## Discussion

Endometriotic lesions are commonly composed of endometriotic glands and stroma surrounded by a variable amount of fibromuscular tissues. The fibromuscular component is considered to be the major cause of pelvic pain and sexual dysfunction in patients with endometriosis [11]. In some lesions, such as rectovaginal endometriosis, the major component is smooth muscle and fibrosis rather than endometriotic tissue. SMM and fibrosis are believed to be ongoing processes that increase with the progression of the disease. The mechanism of SMM formation in endometriotic lesions remains unknown. According to the “Induction

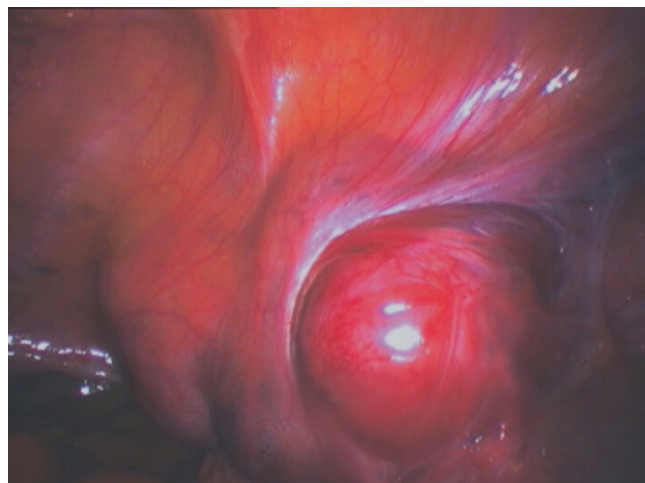


Fig. 2. In case 2, laparoscopy shows a right-sided pelvic mass at the location of the previous right ovary.

theory” as proposed by Levander and Normann in 1955 [12] or the “subcoelomic mesenchyme transformation theory,” the multipotent cells of the peritoneal mesothelium and underlying mesenchyme in the pelvis and lower abdomen are considered to be the “secondary Müllerian system” because of their embryologic resemblance to the Müllerian ducts [13]. During embryogenesis, the primitive pelvic coelomic cells are the source of the urogenital tissue that encompasses the Müllerian and Wolffian ducts [3]. These cells have the ability to convert into uterine tissue by differentiating into endometrial stromal cells, decidua or smooth muscle cells under hormonal influences. Coelomic cells undergoing SMM are hormone-receptive. The cells have been found to have estrogen and progesterone receptors, as well as oxytocin receptors [14,15].

These 2 case reports postulate different pathogenic mechanisms leading to the formation of uterus-like endometriotic mass. In the first case, the mass appeared to be a straightforward case of a noncommunicating rudimentary uterine horn in which endometriosis may have arisen either from retrograde menstruation and dysperistalsis from an early obstructive defect in the primary Müllerian system or from a defect in the secondary Müllerian system, which had undergone smooth muscle metaplasia. In the second case, the uterus-like mass may have arisen from either an endometriotic ovarian remnant left from previous surgery, which underwent SMM under the influence of the unopposed estrogen that the patient had been receiving, or an undiagnosed Müllerian remnant that had grown under hormonal influences.

The “uterus-like mass” may be the end stage of transition from ovarian stromal cells or endometriosis cells into smooth muscle cells, because of local metaplasia under the influences of endometriotic tissue hormonal secretions. Another possibility is that the “uterus-like mass” is a remnant of a congenital Müllerian fusion defect. It has been shown that Müllerian anomalies are strongly related to endometriosis [16]. When the association was demonstrated in obstructive Müllerian anomalies, it was presumed that the pathophysiology is based on retrograde menstruation. Further studies demonstrated a high rate of endometriosis in patients with nonobstructive anomalies, possibly as a result of uterine dysperistalsis. But ovarian endometriosis and findings resembling myomas or adenomyosis have also been found in patients with Mayer-Rokitansky-Küster-Hauser syndrome [17–22] and in premenarcheal girls [23], hinting more toward a Müllerian defect in both the primary and secondary Müllerian systems.

Uterus-like masses have been found in the scrotums of men receiving estrogen therapy for prostatic carcinoma as a result of “secondary Müllerian system” transformation [24]. It can also be found in other parts of the body because of migration defects or lymphatic spread. Ovarian stromal SMM can be seen mostly in the theca externa and cortical stroma of the ovary [10]. It is associated with folliculogenesis and is presumed to provide smooth muscle contractility

to the perifollicular ovarian stroma for ovulation. SMM has also been found to a greater extent in patients with endometriosis and other pathologic sequences including hyperthecosis, granulosa-stromal cell tumors, cystic epithelial ovarian neoplasms and leiomyomatosis peritonealis disseminata [8,10]. Other ovarian stromal metaplasias include ovarian decidualization, as well as adipose tissue and bone metaplasia [8]. SMM associated with endometriosis can result from either metaplastic endometriotic multipotent stromal cells within the ovary or metaplastic ovarian stromal cells under local hormonal influences.

The uterus-like mass may be the end result of a defect in either the primary or secondary Müllerian systems or it can be a single defect affecting both systems. Finding the answer to that may shed light on the complex pathogenesis of endometriosis.

## References

1. Cozzutto C. Uterus-like mass replacing ovary: a report of a new entity. *Arch Pathol Lab Med.* 1981;105:508–511.
2. Pueblitz-Peredo S, Luévano-Flores E, Rincón-Taracena R, Ochoa-Carrillo FJ. Uterus-like mass of the ovary: endomyometiosis or congenital malformation? A case with a discussion of histogenesis. *Arch Pathol Lab Med.* 1985;109:361–364.
3. Redman R, Wilkinson EJ, Massoll NA. Uterine-like mass with features of an extrauterine adenomyoma presenting 22 years after total abdominal hysterectomy-bilateral salpingo-oophorectomy. *Arch Pathol Lab Med.* 2005;129:1041–1043.
4. Rahilly MA, Al-Nafussi A. Uterus-like mass of the ovary associated with endometrioid carcinoma. *Histopathology.* 1991;18:549–551.
5. Ahmed AA, Swan RW, Owen A, Kraus FT, Patrick F. Uterus-like mass arising in the broad ligament: A metaplasia or Müllerian duct anomaly? *Int J Gynecol Pathol.* 1993;16279–281.
6. Mitra S, Nicol A, Scott GI. Uterus-like mass of the ovary. *J Obstet Gynaecol.* 1997;17:94–95.
7. Shutter J. Uterus-like ovarian mass presenting near menarche. *Int J Gynecol Pathol.* 2005;24:382–384.
8. Fukunaga M. Smooth muscle metaplasia in ovarian endometriosis. *Histopathology.* 2000;36:348–352.
9. Rohlfing MB, Kao KJ, Woodward BH, Durham NC. Endometriosis: possible association with leiomyomatosis disseminate and endometriosis. *Arch Pathol Lab Med.* 1981;105:556–557.
10. Doss BJ, Wanek SM, Jacques SM, Qureshi F, Ramirez NC, Lawrence WD. Ovarian smooth muscle metaplasia: An uncommon and possibly unrecognized entity. *Int J Gynecol Pathol.* 1999;18:58–62.
11. Itoga T, Matsumoto T, Takeuchi H, et al. Fibrosis and smooth muscle metaplasia in rectovaginal endometriosis. *Pathol Int.* 2003;53:371–375.
12. Levander G, Normann P. The pathogenesis of endometriosis; and experimental study. *Acta Obstet Gynecol Scand.* 1955;34:366–398.
13. Lauchlan S. The secondary Mullerian system. *Obstet Gynecol Surv.* 1972;27:133–46.
14. Kitano T, Matsumoto T, Takeuchi H, et al. Expression of estrogen and progesterone receptors in smooth muscle metaplasia of rectovaginal endometriosis. *Int J Gynecol Pathol.* 2007;26:124–129.
15. Mechsner S, Bartley J, Loddenkemper C, Salomon DS, Starzinski-Powitz A, Ebert AD. Oxytocin receptor expression in smooth muscle cells of peritoneal endometriotic lesions and ovarian endometriotic cysts. *Fertil Steril.* 2005;83:1220–1231.
16. Nawroth F, Rahimi G, Nawroth C, Foth D, Ludwig M, Schmidt T. Is there an association between septate uterus and endometriosis? *Hum Reprod.* 2006;21:542–544.

17. Kaya H, Sezik M, Ozkaya O, Sahiner H. Large endometrioma in an adolescent girl with Mayer-Rokitansky-Küster-Hauser syndrome. *Gynecol Surg.* 2004;1:241–242.
18. Al-Fadhli R, Tulandi T. A rare case of completely separated rudimentary uterine horns with myoma and adenomyosis. *J Am Assoc Gynecol Laparosc.* 2006;13:86–87.
19. Jadoul P, Pirard C, Squifflet J, Smets M, Donnez J. Pelvic mass in a woman with Mayer-Rokitansky-Kuster-Hauser syndrome. *Fertil Steril.* 2004;81:203–204.
20. Enatsu A, Harada T, Yoshida S, Iwabe T, Terakawa N. Adenomyosis in a patient with the Rokitansky-Kuster-Hauser syndrome. *Fertil Steril.* 2000;73:862–863.
21. Tsin DA, Waters TK, Granato RC. Laparoscopic myomectomy in a patient with Mayer-Rokitansky-Kuster-Hauser syndrome. *J Am Assoc Gynecol Laparosc.* 2000;7:411–413.
22. Deligeorgiou E, Kontoravdis A, Makrakis E, Christopoulos P, Kountouris A, Creatas G. Development of leiomyomas on the uterine remnants of two women with Mayer-Rokitansky-Kuster-Hauser syndrome. *Fertil Steril.* 2004;81:1385–1387.
23. Marsh EE, Laufer MR. Endometriosis in premenarcheal girls who do not have an associated obstructive anomaly. *Fertil Steril.* 2005;83:758–760.
24. Scully RE. Smooth-muscle differentiation in genital tract disorders. *Arch Pathol Lab Med.* 1981;105:505–507.