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.

**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 several myomas in the left horn and a small right horn distended with blood and bilateral endometriomas associated with a normal right kidney and a left hypertrophied kidney with normal excretion of the contrast agent. Cystoscopy showed an absent right ureteric orifice. During laparoscopy the right pelvic sidewall and retroperitoneum were dissected. The
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-
theory” as proposed by Levander and Normann in 1955 [12]
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 coe-
locic 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 dif-
ferentiating 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 recep-
tors, as well as oxytocin receptors [14,15].

These 2 case reports postulate different pathogenic
mechanisms leading to the formation of uterus-like endo-
metriotic 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 uterine-like mass may have arisen from either an endo-
metriotic ovarian remnant left from previous surgery, which
underwent SMM under the influence of the unopposed es-
tragen the patient had been receiving, or an undiag-
nosed 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. An-
other 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 endometri-
osis [16]. When the association was demonstrated in ob-
structive Müllerian anomalies, it was presumed that the
pathophysiology is based on retrograde menstruation. Fur-
ther 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 premenarchal 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 folliculogen-
esis 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 endo-
metriosis and other pathologic sequences including hyper-
theosis, granulosa-stromal cell tumors, cystic epithelial
ovarian neoplasms and leiomyomatosis peritonealis dis-
seminata [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 stro-
mal 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 an-
swer 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
2. Pueblitz-Peredo S, Luévano-Flores E, Rincón-Taracena R, Ochoa-
Carrillo FJ. Uterus-like mass of the ovary: endomyometriosis or con-
genital malformation? A case with a discussion of histogenesis. Arch
3. Redman R, Wilkinson DJ, Massoll NA. Uterine-like mass with fea-
tures of an extraterine adenomyoma presenting 22 years after total
abdominal hysterectomy-bilateral salpingo-oophrectomy. Arch
4. Rahilly MA, Al-Nafussi A. Uterus-like mass of the ovary associated
5. Ahmed AA, Swan RW, Owen A, Kraus FT, Patrick F. Uterus-like
mass arising in the broad ligament: A metaplasia or Müllerian duct
7. Shutter J. Uterus-like ovarian mass presenting near menarche. Int J
9. Rohlfing MB, Kao KJ, Woodward BH, Durham NC. Endometriosis:
possible association with leiomyomatosis disseminate and endo-
10. Doss BJ, Wanek SM, Jacques SM, Qureshi F, Ramirez NC, Lawrence
WD. Ovarian smooth muscle metaplasia: An uncommon and possibly
12. Leverand G, Normann P. The pathogenesis of endometriosis: and
1972;27:133–46.
progesterone receptors in smooth muscle metaplasia of rectovaginal
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
there an association between septate uterus and endometriosis? Hum


