

# Uterus-Like Mass with Features of an Extrauterine Adenomyoma: A Case Report and Literature Review

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Uterus-like masses, such as cavities lined by endometrium-type mucosa surrounded by bundles of smooth muscle cells, may strikingly resemble the uterus. In this report, we describe a case of a uterus-like mass with features of an extrauterine adenomyoma in a 42-year-old woman. The first uterine-like mass was documented by Cozzutto in 1981 and to date, 13 such cases have been reported. Three theories have been offered to explain their etiology: 1) the uterine/Mullerian duct fusion defect theory, which is based on a developmental abnormality occurring during the formation of the female genital tract, 2) the metaplastic theory, which is based on the fact that a uterus-like mass may arise from subperitoneal mesenchymal cells that retain the ability to duplicate Mullerian duct structures, and 3) the heterotopia theory. We consider that the metaplastic theory best fits with our observations in the present case as both glandular and stromal smooth muscle cells proliferated as a true neoplasm rather than as an anomaly.

**Key Words** : Adenomyoma; Endometriosis; Pelvic region

Uterus-like masses are composed of endometrial tissue and smooth muscle, and histologically resemble the uterus. Most arise within the ovary, but extraovarian cases have been described. Cozzutto in 1981 suggested that this lesion originates from ovarian stromal cells that have undergone smooth muscle cell metaplasia during endometriosis.<sup>1</sup> To date, 13 cases of uterine-like masses have been reported in the English literature, but the histogenesis of these masses is controversial. In this report, we report a case of a uterus-like mass in the pelvic cavity, and review the relevant literature.

## CASE REPORT

A 42-year-old woman complained of lower abdominal pain of 4 days duration. Pelvic computed tomography (CT) at a local clinic revealed a well defined lobulated cystic mass with multiple septations in the pelvic cavity. The obstetrical history of this patient was 3 gravida, 2 parity, 1 artificial abortion, and 2 cesarean sections. Her serum CA125 level was elevated (269.09 U/mL). At surgery, a 10.5 × 9.5 cm sized mass was located in periaxonal area between the sigmoid colon and posterior fundus of the uterus with attachment to these organs. The mass

was grossly well circumscribed and had variably sized cystic spaces surrounded by a thick layer of smooth muscle. The cystic portion looked like endometrium and contained dark brown viscous material, and its inner surface was coated by multiple adherent blood clots.

Microscopically, the mass was composed of a thick muscular cyst wall lined with benign endometrial glands and endometrial stroma with an arrangement resembling endometrium (Fig. 1). The endometrial glands had proliferative to hyperplastic features (Fig. 2A, B) and the cystic wall consisted of thickened smooth muscle bundles that resembled myometrium (Fig. 2C).

Immunohistochemical analysis was performed on 10% formalin-fixed, paraffin-embedded sections using the following antibodies; cytokeratin (AE1/AE3) (1:50, DAKO, Glostrup, Denmark), epithelial membrane antigen (EMA) (E29, 1:60, DAKO, Glostrup, Denmark), antihuman mesothelial cell (HBME-1) (1:40, DAKO, Glostrup, Denmark), S-100 (1:150, Zymed, San Francisco, CA, USA), vimentin (1:150, Zymed, San Francisco, CA, USA), desmin (ZC18, 1:60, Zymed, San Francisco, CA, USA), smooth muscle actin (SMA) (1A4, 1:40, DAKO, Glostrup, Denmark), estrogen receptor (ER) (1:50, Zymed, San Francisco, CA, USA), and progesterone receptor (PR) (1:60, Zymed, San Francisco, CA, USA). The endometrial glandular

cells were positive for cytokeratin (AE1/AE3), EMA, HBME-1, ER, and PR, endometrial stromal cells were positive for SMA, vimentin, HBME-1, ER, and PR and myometrial components were positive for SMA, desmin, vimentin, HBME-1, ER, and PR (Table 1, Fig. 3).

Based on these findings, the lesion was diagnosed as a uterus-like mass.

## DISCUSSION

Uterus-like masses are a rare entity of uncertain histogenesis.

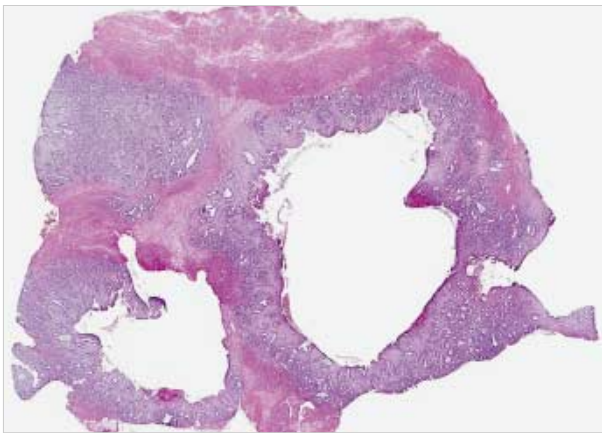


Fig. 1. The periadnexal mass showing a cavity lined by endometrium-type mucosa surrounded by bundles of smooth muscle cells, resembling the uterus.

Most arise within the ovary, but extraovarian cases have been described. Uterus-like masses should be differentiated from adenomyomas, which lack a uterus-like organization. The first uterine-like mass was documented by Cozzutto in 1981. He described a mass originating from stromal cells due to smooth muscle cell metaplasia, possibly due to the effect of estrogen. Fibroblasts and smooth muscle cells appear to be closely related to myofibroblasts, and possibly participate in the transitional stage.<sup>1</sup> To date, 13 cases of uterus-like mass have been reported (Table 2).<sup>1-11</sup> A review of these cases and the present case showed that all occurred in women aged 11 to 59 years (average age 35 years). Mass sizes ranged from 2.5 to 16 cm with an average of 8 cm. Nine cases presented with complaints of lower abdomi-

Table 1. Immunohistochemical findings of periadnexal mass

Antibody	Reactivity		
	Endometrial glandular cells	Endometrial stromal cells	Myometrial component
Cytokeratin (AE1/AE3)	+	-	-
Epithelial membrane antigen (EMA)	+	-	-
Antihuman mesothelial cell (HBME-1)	+	+	+
Vimentin	-	+	+
Smooth muscle actin (SMA)	-	+	+
Desmin	-	-	+
S-100	-	-	-
Estrogen receptor (ER)	+	+	+
Progesterone receptor (PR)	+	+	+

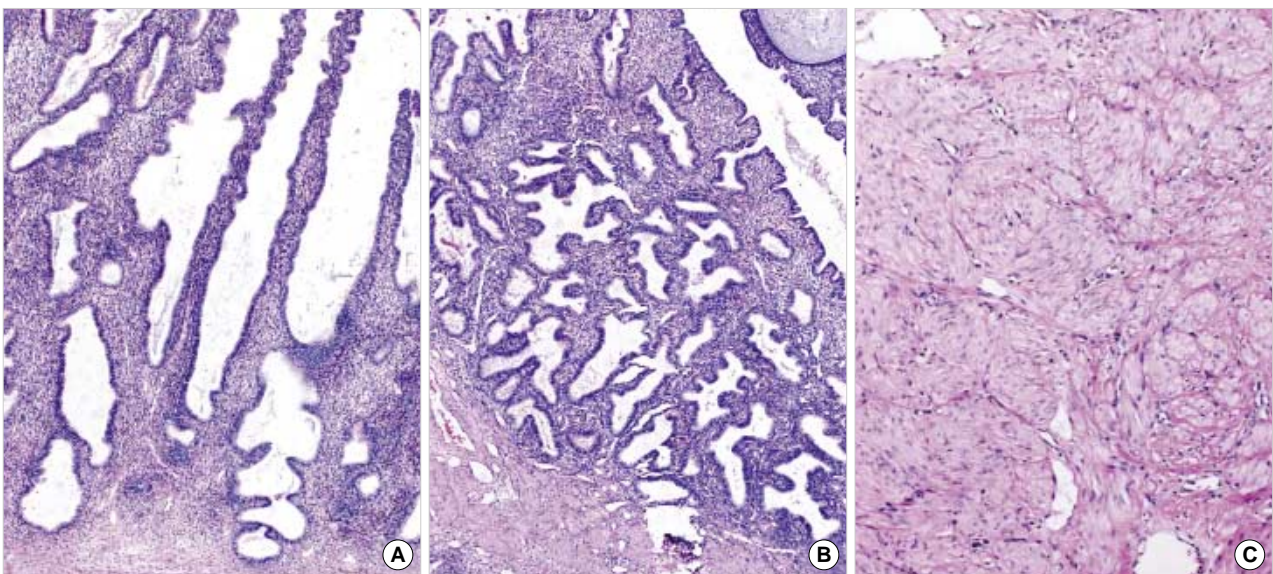


Fig. 2. (A) Proliferative, (B) hyperplastic endometrial glands, and (C) thickened smooth muscle bundles appearing similar to myometrium ( $\times 100$ , H&E stain).

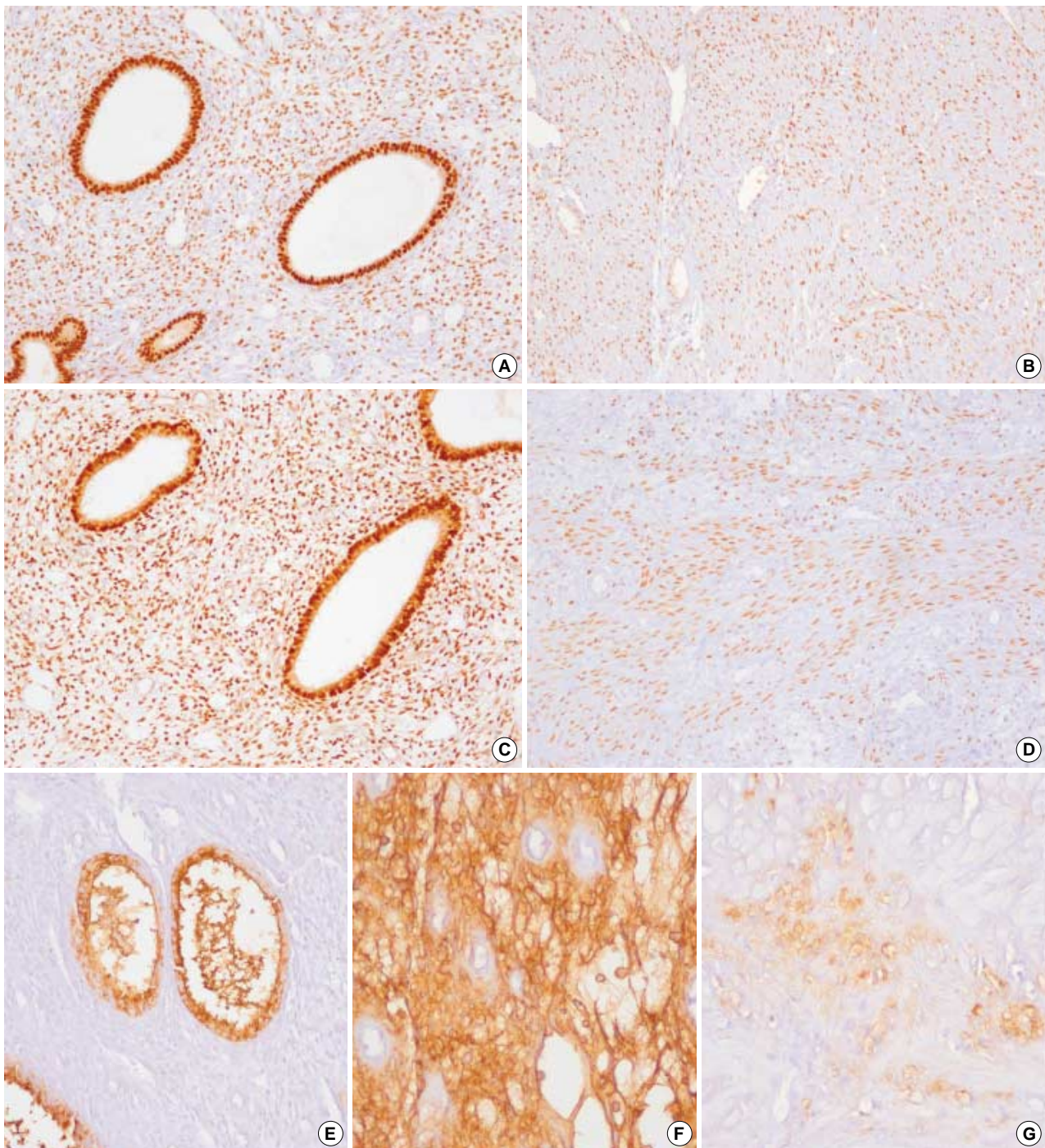


Fig. 3. Endometrial components, glandular and stromal cells, and myometrial components are immunoreactive for ER (A, B), PR (C, D), and HBME-1 (E, F, G).

nal pain with/without bleeding, three cases were found incidentally during surgery or follow-up, one case presented as a palpable mass, and another presented with paresthesia and weakness of the legs. All 14 cases involved thick muscular walled masses with a central cavity or multicystic space, resembling a uterus.

Eight cases arose from the ovary, and six cases from extraovarian regions, including the broad ligament, the small intestine, the small bowel mesentery, the conus medullaris, and the periaxonal region. To our knowledge, the present lesion is the first case of a uterus-like mass arising from the periaxonal region.

**Table 2.** Summary of reported cases of uterus-like mass in the literature

Case No. (reference)	Age/sex	Size (cm)	Location	Associated finding	Theory	Clinical symptom	Gross finding
1 (1)	31/F	6	Left ovary	Left renal agenesis	Metaplasia	Pelvic pain	Round, smooth surfaced mass with central, round cavity
2 (2)	18/F	9	Right ovary	Double excretory system	Anomaly	Pelvic pain	Thick-walled mass with central cavity
3 (3)	12/F	2.5 and 1.2	Small intestine	Anomalies fo the lower genital and itestinal tracts, sacral agenesis, and a presental or sacrococcygeal teratoma	Heterotopia	Incidental finding of surgery	Compact concentric smooth muscle surrounding a central cavity
4 (4)	38/F	9	Right ovary	Endometrioid carcinoma in same ovary and uterus	Metaplasia	Pelvic pain	Thick muscular wall with a central cavity
5 (5)	18/F	2.5	Conus medullaris	Spinal dysraphism, lipoma	Anomaly	Paresthesia and weakness of leg	Ovoid mass with central cavity
6 (6)	49/F	8	Right ovary	Clonal chromosome deletion 2p21	Metaplasia	Lower abdominal pain and menorrhagia	Solid mass with a round central cavity containing dark, chocolate like liquid
7 (7)	46/F	16	Right broad ligament	Third fallopian tube	Metaplasia	Abdominal pain and bleeding	Encapsulated mass with multiloculated cyst containing serosanguinous fluid
8 (8)	38/F	4	Left ovary	Breast carcinoma	Metaplasia	Incidental finding during breast cancer follow up	Thick-walled cyst filled with chocolate brown grumous material, resembling a uterus with a cavity
9 (8)	43/F	11.5	Ovary (side unknown)	Elevated CA125	?	Incidental finding of surgery	Thick-walled mass with central cyst
10 (8)	39/F	13	Left ovary	Breast carcinoma, elevated CA125	Metaplasia	Lower abdominal pain	Thick-walled mass with central cyst resembling a uterus
11 (9)	59/F	14	Small bowl mesentery	Uterine lipoleiomyoma	Metaplasia	Palpable mass	Well-demarcated mass with multicystic spaces
12 (10)	50/F	5	vaginal cuff	History of total abdominal hysterectoy and bilateral salpingo-oophorectomy	Metaplasia	Dysuria, suprapubic and pelvic pain	Pear-shaped mass with a cystic center
13 (11)	11/F	4.5	Right ovary	Chronic vesicoureteral reflux	Anomaly	Lower abdominal pain	Smooth-lined cystic mass with a thick wall
Present case	42/F	10.5	Periadnexa	Elevated CA125	Metaplasia	Lower abdominal pain	Thick-walled mass with multicystic spaces

Three hypotheses for the histogenesis of these masses have been proposed: 1) Mullerian duct fusion defect, 2) metaplasia, and 3) heterotopia theories. The Mullerian duct defect theory is based on a developmental abnormality during the formation of the female genital tract. Male and female embryos have two pairs of genital ducts the Wolffian (mesonephric) duct and the Mullerian (paramesonephric) duct. The Mullerian duct begins as a longitudinal folding of the coelomic epithelium on the antelateral surface of the urogenital ridge and becomes the main female genital duct. With ovary descent at gestation week 9,

the uterine tube and uterine canal are formed by the fusion of three separate portions of the Mullerian duct and extended cranially to caudally. Lack of fusion of the Mullerian duct in a localized area, or throughout its length, may explain various duplications or atresias of the uterus.<sup>12</sup> Uterus-like masses may result from either a Mullerian duct fusion defect or a true partial duplication of the Mullerian system, and represent uterine tissue that is anatomically separated from the uterine corpus; moreover, they are probably an example of uterus unicornis.<sup>13</sup>

The second metaplastic theory is based on the finding that

uterus-like masses may arise from subperitoneal mesenchymal cells that retain the ability to duplicate Mullerian duct structures. In 1997, it was proposed that some uterus-like masses originate from the peritoneal mesothelium and its subjacent connective tissue. These tissues probably retain the potential to produce different Mullerian structures, like the uterus and fallopian tubes.<sup>7</sup> Moreover Lauchlan<sup>14</sup> once proposed the concept of a “secondary Mullerian system” composed of peritoneal or retroperitoneal tissues with the potential to differentiate into cells similar or identical to those lining the oviducts, the endometrial cavity, or the endocervix. Support for the hormonal responsiveness of the secondary Mullerian system is provided by the uterus-like masses observed in the scrotums of men receiving estrogen therapy for prostate carcinoma.<sup>15</sup> Pai *et al.*<sup>8</sup> found a close relationship between ovarian uterus-like masses, breast cancer, and elevated serum CA125 levels, and concluded that uterus-like masses are a hormone-dependent lesions and a form of endometriosis. A case was also reported in which endometriosis of a pelvic lymph node was associated with an adjacent nodular proliferation of smooth muscle cells, which the authors called “endomyometriosis”.<sup>16</sup>

The third heterotopia theory was proposed by Peterson *et al.*<sup>3</sup> who reported a case of an ileal uterus-like mass in a 12-year-old girl associated with multiple lower intestinal and urogenital tract anomalies and a history of sacrococcygeal teratoma. This mass bore a striking resemblance to the uterine fundus and fallopian tube. Accordingly, they concluded that neither the congenital anomaly nor the metaplasia theory provided an entirely satisfactory explanation regarding the causative effects of either heterotopia or choristoma.

In our case, the patient had neither a structural uterine abnormality consistent with a Mullerian fusion defect nor congenital renal abnormalities on extensive workup, which included CT, ultrasound and an intravenous pyelogram. By immunohistochemistry, endometrial components, glandular and stromal cells, and myometrial components of the mass were positive for HBME-1, PR, and ER. Positivity for HBME-1 suggests that the mass originated from peritoneal mesothelial cells, i.e., “secondary Mullerian system”, and positivity for ER and PR suggests that the mass was responsive to hormonal stimulation. In fact, the patient had an elevated serum CA125 levels (269.09 U/mL).

In conclusion, we consider that the metaplastic theory best fits with our observations in the present case as both glandular and stromal smooth muscle cells proliferated as a true neoplasm rather than as an anomaly.

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