Uterine-like Mass With Features of an Extrauterine Adenomyoma Presenting 22 Years After Total Abdominal Hysterectomy—Bilateral Salpingo-oophorectomy

A Case Report and Review of the Literature

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Adenomyoma is a benign tumor composed of smooth muscle and benign endometrium. These tumors typically originate within the uterus. An extrauterine adenomyoma is a rare entity. We report a uterine-like mass consistent with an extrauterine adenomyoma presenting 22 years following a total abdominal hysterectomy and bilateral salpingo-oophorectomy. The mass was pear-shaped with uterine-type smooth muscle and a cavity lined by functional endometrial glands and stroma. To our knowledge, only 4 other cases of an extrauterine uterine-like mass are reported in the literature. Three involved the ovary, while one was located adjacent to the broad ligament with normal pelvic organs. Although none of these other uterus-like masses were described as adenomyomas with uterine-like features, the histologic findings are strikingly similar. An understanding of the müllerian system suggests that either an embryologic malformation or a differential multipotentiality existing in the subcoelomic tissues in response to hormonal stimulation results in a supernumerary müllerian structure like a uterus, as observed in this case. The presence of endometrial glands and stroma in the mass confirms that the tissues in this mass are hormonally responsive. It is most likely that this uterine-like mass arose from the tissues of the secondary müllerian system in response to estrogenic stimulation.

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Extraperitoneal-like masses within the pelvis may arise from the uterus, from within the broad ligament, from the fallopian tube, or from the ovary. Leiomyomas arising from uterine smooth muscle account for a small percentage of the cases. Adenomyomas, benign tumors composed of smooth muscle and nonneoplastic endometrium, typically originate within the uterus. An adenomyoma presenting outside the uterus is uncommon. Six cases of uterine-like masses, exterior to the uterus, have been reported.1–5 Of these, 2 have been described as adenomyoma with uterus-like features.1 This report documents a third case of a uterus-like mass with features of an adenomyoma. A review of the literature, to further understand the frequency and etiology of this unusual finding, is presented.

REPORT OF A CASE

The patient was a 50-year-old woman who presented with complaints of dysuria and suprapubic and pelvic pain. She was treated for 7 months prior to surgery for recurrent urinary tract infections. Her medical history was significant for a total abdominal hysterectomy and bilateral salpingo-oophorectomy 22 years earlier for benign disease. Medications included regular use of an estrogen patch, aspirin, and vitamins. A pelvic examination revealed a palpable 8.0-cm, firm, irregular mass at the vaginal cuff pressing on the rectum and extending into the obturator space. Workup included an intravenous pyelogram that showed a left ureteral stricture at the ureterovesical junction. A computed tomographic scan and gynecologic ultrasound showed a large, heterogeneous, solid pelvic mass on the left side of the vaginal cuff with associated left hydroureteronephrosis. A ureteral stent was placed to relieve the obstruction.

At laparotomy, a pelvic mass measuring 8.0 cm in greatest dimension was found that partially encased the distal left ureter at the level of the pelvic brim. The mass was surgically excised, requiring marsupilization for complete excision. On gross examination, the mass appeared encapsulated; a portion of the mass was pear-shaped, measuring 5.0 × 5.0 × 4.0 cm, with a cystic center. Upon sectioning the cystic portion, approximately 50 mL of dark brown viscous fluid exuded, revealing a firm white cyst lining and multiple adherent blood clots (Figure 1). A frozen section was performed on the cystic portion of the mass and interpreted as endometriosis.

A microscopic evaluation showed a thick muscular cyst wall (Figure 2, a) lined with endometrial stroma and glands (Figure 2, b). The endometrial glands had secretory features as well as morulae metaplasia. A prominent smooth muscle component, reminiscent of uterine wall myometrium, accounted for most of the cyst wall. The mass had a serosal surface with some fibrovascular adhesions. Foci of endometrium, consistent with adenomyosis, were present within the smooth muscle. Variable amounts of blood, necrotic-appearing endometrium, and hemosiderin-laden macrophages were present throughout the mass.
The differential diagnosis included endometrioma, leiomyomatosis peritonealis disseminata, and extracellular adenomyoma. The prominent smooth muscle component distinguished this mass from an endometrioma. Leiomyomatosis peritonealis disseminata was also excluded because of the presence of cycling endometrial tissue and the lack of a recent or current pregnancy. The mass was diagnosed as an extracellular adenomyoma with uterine-like features.

**COMMENT**

Six cases of an extracellular uterine-like mass have been reported in the literature, of which 2 were described as a uterine adenomyoma with uterine-like features. Three masses involved the ovary, and one was adjacent to the broad ligament, with otherwise normal pelvic organs. Sizes ranged from 6 to 16 cm. The gross appearance, including cystic and solid areas, varied between cases. The microscopic descriptions of each mass included functional endometrial glands and stroma within a muscular wall resembling a uterus.

Two theories have been offered to explain the etiology of the uterine-like mass: the uterine/mullerian duct fusion defect theory and the subcoelomic mesenchyme transformation theory. In a 1982 letter to the editor in the Archives of Pathology & Laboratory Medicine, Rosai suggested a theory as to the origin of the first uterine-like mass documented by Cozzutto in 1981. Cozzutto postulated that the mass originated from ovarian stromal cells that underwent smooth muscle cell metaplasia in a focus of endometriosis, possibly through a stage of myofibroblasts. Cozzutto described the mass as an ovarian leiomyoma on the basis of its ultrastructural features. Electron microscopy of the wall of the cyst showed a predominance of elongated smooth muscle cells as well as cells consistent with myofibroblasts. Cozzutto later revised the diagnosis of leiomyoma. He thought the mass was more like a uterus than a leiomyoma. Rosai suggested that the mass was uterine-like because it was related to the uterus, not to the ovary, and postulated that the lesion resulted from a mullerian duct fusion defect.

The mullerian duct fusion defect theory is based on a developmental abnormality occurring during the formation of the female genital tract. Initially, male and female embryos have 2 pairs of genital ducts: Wolffian (mesonephric) and mullerian (paramesonephric). The mullerian duct begins as a longitudinal folding of the coelomic epithelium on the anterolateral surface of the urogenital ridge and becomes the main genital duct of the female. With the descent of the ovary at 9 weeks' gestation, the uterine tube and uterine canal are formed from the fusion of 3 separate portions of the duct extending cranially to caudally. Lack of fusion of the mullerian ducts in a localized area, or throughout the length of the ducts, may explain various duplications or atresias of the uterus. Of the many anatomic variants of mullerian duct fusion defects, Rosai proposed that a well-formed uterus with a single horn, the uterus unicorns, can be associated with a rudimentary detached uterine horn in the contralateral adnexal region, providing the most plausible explanation for this mass.

Pueblitz-Peredo et al published the second case report of uterine-like masses and postulated that the masses were the result of a mullerian duct fusion defect. They noted,
however, that the rudimentary horn concept of a uterus unicornis was not sufficient to account for the relatively large uterus-like mass found. These authors proposed that the malformation was a “rudimentary horn,” in which a rudimentary müllerian structure with a central cavity lies in an adnexal position to the well-developed uterus with the exterior surfaces connecting via fibrous tissue.

The subcoelomic mesenchymal transformation theory challenges the müllerian fusion defect theory. The subcoelomic mesenchyme is defined as the layer of tissue that lies underneath the mesothelial surface of the peritoneum. In the primitive pelvic coelom, this layer of tissue gives rise to the mesenchyme of the urogenital ridge that surrounds the early müllerian and wolffian ducts. In the adult, the subcoelomic mesenchyme or secondary müllerian system is represented by “…an inconspicuous layer of flattened cells that blend imperceptibly into the subserosal stroma of the uterus, ovaries, tubes, and uterine ligaments.” While not technically part of the female genital tract, the cells of the secondary müllerian system are thought to be multipotential and may proliferate in response to hormonal stimulation. Epithelial lesions of this system may show normal or neoplastic endometrioid, serous, mucinous, or even transitional differentiation. Proliferation of the subjacent mesenchyme may give rise to mesenchymal lesions composed of endometrial stromal-type cells, decidua, or smooth muscle. Support for the hormonal responsiveness of the secondary müllerian system is given by the uterus-like masses observed in the scrotums of men receiving estrogen therapy for prostate carcinoma, as described by Scully. Displacement of coelomic epithelium and subcoelomic mesenchyme during embryonic development could account for müllerian tissues or lesions of these tissues elsewhere in the body, including the peripheral sinuses of lymph nodes. Rholfing et al described a pelvic lymph node containing endometrial glands and stroma surrounded by smooth muscle within the central and peripheral sinuses of the lymph node, a finding they called endomyometriosis. This lesion was believed to be morphologically distinct from either endometriosis or leiomyomatosis peritonealis disseminata because of its simulation of a uterus.

Like the endomyometriosis of the lymph nodes described by Rholfing et al, the histologic changes observed in this case recapitulate a uterus. However, unlike the findings in the case of Rholfing et al, the mass itself has organization and is not part of an existing structure, like a lymph node. Therefore, a description of our findings as endomyometriosis is not complete. The presence of a central cavity lined by cycling endometrium surrounded by smooth muscle external from a uterus is unusual for an adenomyoma; however, it is the best descriptor for the entity observed in this study.

In this case, it is unlikely that the patient had a structural uterine abnormality consistent with a müllerian fusion defect, because she had undergone a total abdominal hysterectomy and bilateral salpingo-oophorectomy 22 years earlier. An extensive workup of her urinary tract infections, which resulted from the ureteral obstruction secondarily to the compression of the ureter at the pelvic brim by the uterine-like mass, did not show any congenital renal abnormalities, such as unilateral renal agenesis, as are common in women with a müllerian fusion defect. The prior surgical excision of the uterus, fallopian tubes, and ovaries in this patient makes this theory unlikely as an etiologic explanation for the origin of the mass.

Many lesions of the female genital tract, both benign and malignant, are thought to be derived from the subcoelomic mesenchyme in response to hormonal stimulation. The patient had been receiving regular estrogen therapy because of her premature menopause. The presence of endometrial glands and stroma in the mass confirms that the tissues in this mass are hormonally responsive. It is most likely that this uterine-like mass arose from the tissues of the secondary müllerian system in response to estrogen stimulation.

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References