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Uterus-like mass: A very rare and elusive entity a case report

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Abstract

Background: Uterus-like mass (ULM) is an extremely rare lesion. Gross morphology of ULM resembling a uterus. It can occur in various organs in the abdominal cavity, even in the spinal cord. The histogenesis of ULM remains uncertain. A number of hypotheses have been proposed including metaplasia, congenital anomaly, and heterotopia theory.

Methods: We describe a case of 43-year-old male presented with a complaint of acute low abdominal pain. Pelvic ultrasound found a large pelvic mass embedded in the broad ligament.

Results: The mass contains a variable thickness smooth muscle layer lined with endometrial glands and stroma which resembling a uterus. Eventually, the patient was diagnosed as ULM by histopathological examination.

Conclusion: Except hypomenorrhea, the patient did not have any other associated abnormalities. We suggest this case supports the metaplasia theory that ULM is a benign mass formed by the proliferation of ectopic endometrial stromal cells or pluripotent mesenchymal cells of the 2nd Müllerian system.

Abbreviation: ULM = uterus-like mass.

Keywords: broad ligament, metaplasia, uterus-like mass

1. Introduction

Uterus-like mass (ULM) is an extremely rare benign tumorous lesion. It has gross morphology resembling a uterus.[1] It presents as a central cavity lined by endometrial tissue and surrounded by thick-walled smooth muscles. It once termed endomyometriosis, ULM with features of an extra-uterine adenomyoma, or ovarian leiomyoma.[2,3] Most of ULM occurred in the ovary,[4] extra-ovarian cases involving the uterus,[5] uterosacral ligament,[3] pelvic wall,[6] small intestine,[7] and small intestine mesentery,[8] but few cases of the broad ligament have been reported. Here, we report a rare case of ULM of the broad ligament in a 43-year-old woman who presented with acute abdominal pain and review the literature to further understand of this unusual condition.

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2. Case presentation

2.1. Patient information

A 43-year-old woman presented with a complaint of acute low abdominal pain and lasted for 2 hours. Three months ago, a pelvic ultrasound at another hospital showed that there was a pelvic mass in left pelvic cavity for hypomenorrhea. There was no associated pain or any other symptom. She denied dyspepsia or bowel habit changes. Her menarche occurred at the age of 12 years old and the menstrual cycles were normal. Her medical history was remarkable with hypertension for 8 years and Hashimoto thyroiditis for 3 years which were managed by regular use of hypotensive drug and levothyroxine sodium. Physical examination revealed a palpable about 7.0×5.0 cm, firm, irregular mass at the left latero-uterine area. Tenderness or rebound tenderness was not noted. Tumor marker levels, including alpha-fetoprotein, carcinoembryonic antigen, carbohydrate antigen 19-9, γ-glutamyl-transferase, and alkaline phosphatase, were within normal ranges. Ultrasound examination at our hospital revealed a 7.0×4.6 cm sized mixed mass with irregular shape on the left side of uterus; patchy anechoic area was noted; and no abnormalities of the bladder, ureters, or kidneys were noted (Fig. 1A).

A preoperative diagnosis of a pelvic mass torsion was offered and laparoscopy exploration was performed. Laparoscopy revealed a large pelvic mass embedded in the posterior leaf of the broad ligament. Adhesion was formed between the left uterine adnexa and the intestinal canal. After careful release of the adhesion, the uterus, fallopian tubes, and bilateral ovaries were found anatomically normal. The mass was removed by laparotomy without complications. Frozen sections of the mass were performed and interpreted as benign tumor. Besides, the posterior wall of the uterus shows a size of 2.2×2.1 cm hypoechoic mass.

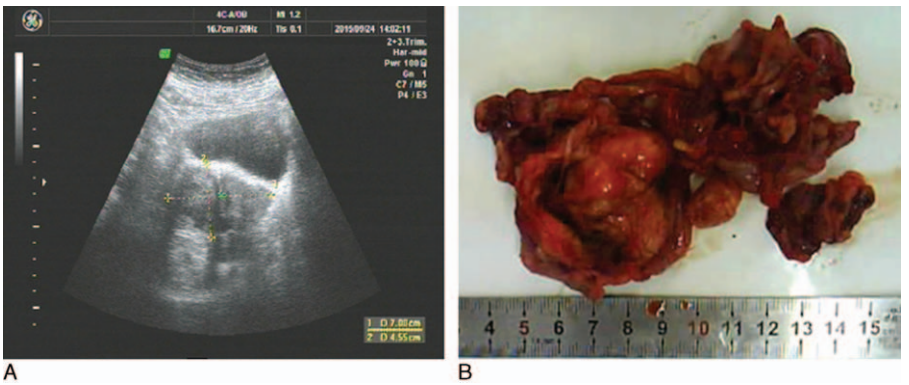

Figure 1. (A) B-mode ultrasonographic finding of a thick-walled cystic mass in the left adnexal region (cross). (B) A well-developed mass with heterogeneous thick layer and central cystic cavity containing dark, chocolate-like liquid.

2.2. Clinical findings

On gross examination, the mass was pear-shaped soft-tissue and well-encapsulated, measuring $7 \times 4.6 \times 4.2$ cm with irregular gray white surface (Fig. 1B). On cut section, the mass showed a central cystic portion which filled with dark red viscous fluid and lined with thin endometrial tissue. On microscopy, a variable thickness of smooth muscle layer lined with endometrial glands and stroma which resembling myometrium and endometrium of a uterus (Fig. 2A). On immunohistochemical staining, the endometrial glandular cells, stromal cells, and smooth muscle cells were positive for estrogen receptor (1:100, Dako, Denmark) (Fig. 2B) and progesterone receptor (1:400, Dako, Denmark) (Fig. 2C). The endometrial stromal cells were also positive for cluster of differentiation 10 (1:100, Dako, Denmark) (Fig. 2D). These findings were consistent with a previously reported case of ULM.[9] All the findings were quite similar to those of a miniature uterus. The final histopathological diagnosis of the mass was consistent with a ULM. In addition, the low echo mass in the posterior wall of the uterus was confirmed to be hysteromyoma.

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.

3. Discussion

A ULM is an extrauterine mass and similar to a normal uterus in structure. It is an extremely rare phenomenon. The ULM was 1st reported in 1981 by Cozzutto.[1] Since then 44 cases have been published and only 5 cases occurred in the broad ligament have

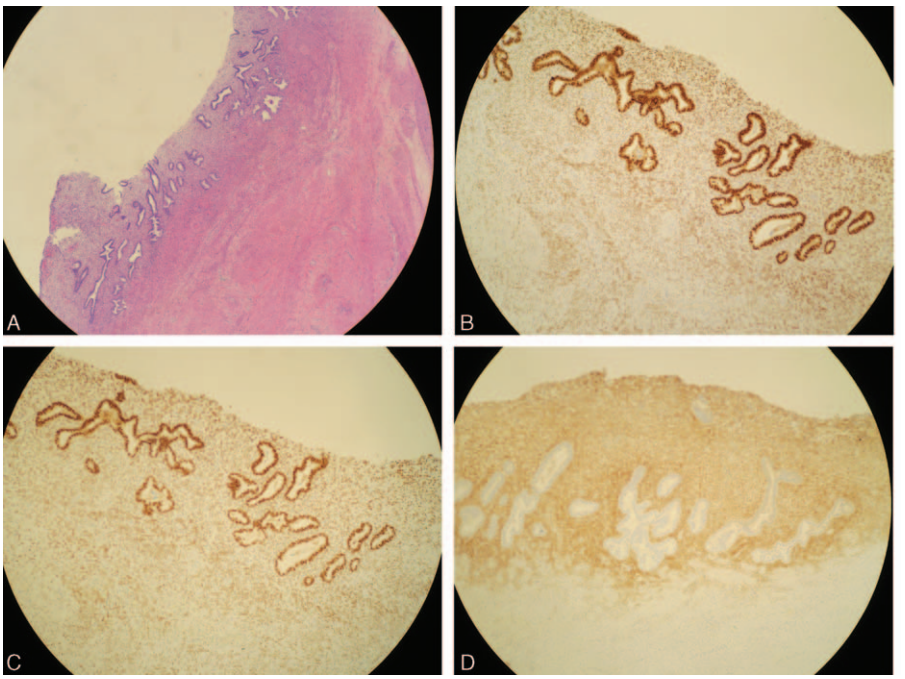

Figure 2. Histological findings of the uterus-like mass. (A) The uterus-like mass showed mature histological structure comprised of stroma and gland overlying smooth muscle (hematoxylin and eosin stain, ×40). (B) Positive estrogen receptor in the nuclei of the epithelial, stromal, and myometrial cells of the uterus-like mass (immunohistochemical stain, ×100). (C) Positive progesterone receptor in the nuclei of the epithelial, stromal, and myometrial cells of the uterus-like mass (immunohistochemical stain, ×100). (D) Positive cluster of differentiation (CD)10 in the cytoplasm of stromal cells of the uterus-like mass (immunohistochemical stain, ×100).

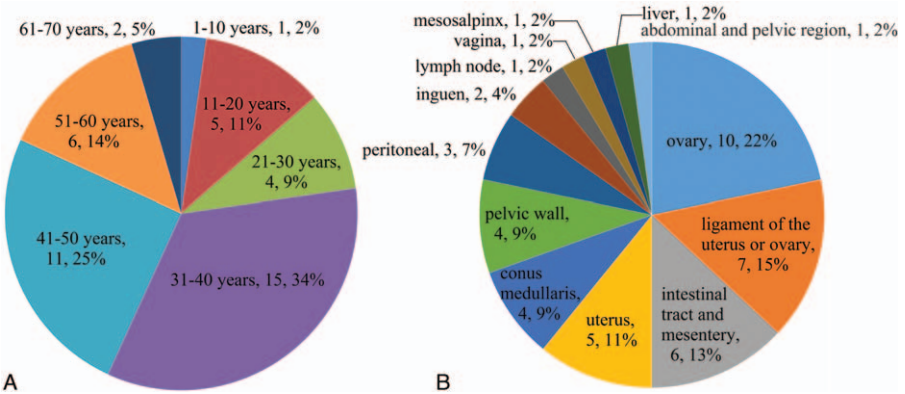

Figure 3. The age (A) and lesional location (B) distribution of previous reported cases of uterus-like mass.

been reported in our review of the literature (Tables 1 and 2). The clinical features of these cases and the present case are summarized in Tables 1 and 2. The ages of the patients ranged from 9 to 67 years and the average age was 38. Three-quarters of the patients were aged between 30 and 60 years old women (Fig. 3A). It is interesting that a man also had a ULM.[24] ULM can occur in various areas, there seems to be some elusive. The ovary is the most common site which followed by ligament of the uterus and ovary. ULM can also occur in the intestinal tract and mesentery, uterus, pelvic wall, peritoneal, inguen, lymph node, vagina, liver, and mesosalpinx, even in the conus medullaris (Fig. 3B). The most common symptom was abdominal pain, frequently associated with dysmenorrhea. Some cases were associated with congenital malformations including spinal split, renal dysplasia, intestinal and urinary tract malformations, and ureteral stricture. A few patients were found by chance. Sizes of the findings of previously reported cases ranged from 2 to 21 cm.[5,16] The masses consistently showed a solid cystic mass. Solid and cystic areas were found in the gross appearance. Histologically, the masses had a well-organized endometrium and myometrium. Similar findings were noted in the present case.

Histogenesis of ULM remains uncertain. A number of hypotheses on the histogenesis of ULM have been proposed including metaplasia, congenital anomaly, and heterotopia theory. The metaplasia theory was generally accepted. Seven similar cases (including 5 cases of the broad ligament, 1 cases of the uterosacral ligament and 1 cases of the ovarian ligament) of tissue of metaplasia origin in association with the ligament of uterus or ovary have been reported previously in the literature (Table 2). It was presumed that UML is transformed by ectopic endometrial stromal cells or pluripotent mesenchymal cells of the 2nd Müllerian system under the stimulation of estrogen.[2,33] The designation "secondary Müllerian system" was proposed by Lauchlan[38] which reference to the pelvic and lower abdominal mesothelium and subjacent mesenchyme with the potential to differentiate into Müllerian type stroma and epithelium. These cells may proliferate in response to hormone stimulation. In adult, the secondary Müllerian system also termed the subcoelomic mesenchyme. It is represented by a layer of flattened cells which merge into the subserosal stroma of uterine ligaments, ovaries, tubes, and uterus. Normal or neoplastic endometrioid, serous, mucinous, or even transitional differentiation may come

Table 1

Age and location features of reported cases of uterus-like mass.

Reference	Age, year	Sex	Lesional location	Reference	Age, year	Sex	Lesional location
Cozzutto/1981[1]	32	F	Ovary	Kaufman and Lam/2008[10]	39	F	Pelvic wall
Rohlfing et al/1981[11]	35	F	Obturator lymph node		57	F	Pelvic wall
Pueblitz-Peredo et al/1985[12]	18	F	Ovary	Carinelli et al/2009[6]	46	F	Pelvic wall and mesentery
Peterson et al/1990[7]	12	F	Ileum		39	F	Ovary and pelvic wall
Rahilly et al/1991[13]	38	F	Ovary	Tijani et al/2010[5]	35	F	Uterus
Rougier et al/1993[14]	18	F	Conus medullaris	Khurana et al/2011[15]	47	F	Abdominal-pelvic region
Mitra et al/1997[16]	34	F	Ovary	Shin et al/2011[17]	31	F	Sigmoid mesocolon
	38	F	Both ovaries	Seki et al/2011[18]	44	F	Inguinal soft tissue
Pai et al/1998[4]	43	F	Ovary		32	F	Pelvic and abdominal peritoneal
	39	F	Ovary	Carvalho et al/2012[19]	41	F	Peritoneal
Horie and Kato/2000[8]	59	F	Ileal mesentery	Kim et al/2012[20]	46	F	Appendix
Fukunaga/2001[21]	47	F	Uterine cervix	Kakkar et al/2012[22]	9	F	Conus medullaris
Jung et al/2002[23]	43	F	Uterus	Na KY et al/2013[9]	39	F	Colon
	52	F	Uterus	González et al/2014[24]	52	M	Inguen
Redman et al/2005[2]	50	F	Vagina	Nakakita et al/2014[25]	67	F	Ovary
Shutter/2005[26]	11	F	Ovary	Li L et al/2014[27]	22	F	Mesosalpinx
Menn et al/2007[28]	37	F	Uterus	Sopha et al/2015[29]	47	F	Liver
Sharma et al/2007[30]	33	F	Conus medullaris	Ko et al/2015[31]	64	F	Pelvic peritoneal
	24	F	Conus medullaris				

Table 2

Clinical features of previously reported cases of uterus-like mass located in ligament of the uterus or ovary.

Reference	Age, year	Sex	Lesional location	Clinical presentation	Associated conditions	Size, cm	Histogenesis
Pai et al/1995[32]	25	F	Broad ligament	Abdominal mass	None	20	None
Ahmed et al/1997[33]	46	F	Broad ligament	Abdominal pain and bloating	None	$16 \times 14 \times 11.5$	Metaplasia
Liang et al/2010[34]	17	F	Broad ligament	Severe dysmenorrhea	Slightly saddle-shaped uterus	4×4	Metaplasia
Bulut et al/2013[35]	56	F	Broad ligament	Menorrhagia, amenorrhea, pelvic, and lumbar pain	Ectopic adrenal tissue	4×1	Metaplasia and Müllerian duct fusion defect
Nechi et al/2013[36]	54	F	Broad ligament	Pelvic pain	Hysterectomy and bilateral		
Adnexectomy	9.3×6.3	None					
Present case	43	F	Broad ligament	Acute abdominal pain	hysteromyoma	$7.0 \times 4.6 \times 4.2$	Metaplasia
Matsuzaki et al/2000[3]	29	F	Uterosacral ligament	Low abdominal pain, dysmenorrhea	None	7.5×7.2	Metaplasia
Takeda et al/2011[37]	39	F	Ovarian ligament	Low abdominal pain	None	$3.8 \times 3.5 \times 3.7$	Metaplasia

into being after the epithelial lesions of this system. The metaplasia theory hold that most of the reported patients were associated with endometriosis but without visceral organ malformations. Kaufman and Lam[10] reported a pelvic ULM which was found in a patient who received estrogen therapy for a long duration after 20 years of the hysterectomy and bilateral salpingo oophorectomy. Scully[39] described that the occurrence of ULM was found in the prostate carcinoma patients who take estrogen therapy. Considering the various locations of ULMs in the reports, it has been speculated that coelomic epithelium and subcoelomic mesenchyme were the sites of origin of the metaplastic cells. Rohlfing et al reported a finding they called endomyometriosis. A pelvic lymph node simulates to a uterus in gross morphology. In the lymph node, the central and peripheral sinuses were consisted of endometrial glands and stroma which were surrounded by smooth muscle.[11]

The congenital anomaly theory suggested that a developmental anomaly leads to ULMs during the formation of the female genital tract. ULMs possibly caused by either a Müllerian duct fusion defects or a partial duplication of the Müllerian system. Urinogenital tract malformation was reported in the literature which often occurred adjacent to the ovary or uterus. This supports the disease associated with embryonic development defects. In 1985, Pueblitz-Peredo et al[12] reported the 2nd case of ULM that was associates with a renal anomaly. The investigators noted the relative size of the ULM was larger than common rudimentary horn. It was not sufficient to clarify this phenomenon with the rudimentary horn concept of a uterus unicornis. They postulated that the mass possibly caused by a Müllerian duct fusion defect. The mass was connected to the exterior surface of the well-developed uterus via fibrous tissue. They proposed that the mass was formed by a rudimentary Müllerian structure which lies in the adnexal position.

The heterotopias theory was proposed by Peterson et al.[7] They found Müllerian duct anomaly and the metaplasia cannot provide an entirely satisfactory explanation for an ileal ULM case with multiple anomalies and concluded that heterotopia or choristoma was the cause. In addition, Rougier et al[14] and Sharma et al[30] reported that ULMs were found in the spinal cords of patients with spina bifida. They suggested that heterotopic Müllerian duct tissue within the neural tube can result in ULMs. Batt[40] proposed the diagnostic criteria for the choristoma which was formed only by a single Müllerian tube: no phenomenon of pelvic endometriosis; no relationship with the endocervix, endometrium, or endosalpinx; and no surgery history on the reproductive organs.

The present case had neither dysmenorrhea nor endometriosis lesions in the pelvic cavity; moreover, the patient had neither uterine anomalies nor other genitourinary anomalies related to Müllerian duct fusion defects; and no previous history of genital tract operation. The mass occurred in the broad ligament. The ectopic endometrial stromal cells or pluripotent mesenchymal cells of the 2nd Müllerian system merge into the subserosal stroma of broad ligament. These cells proliferated and differentiate into endometrium or smooth muscle under the stimulation of estrogen and thus form a uterine-like mass. So, the present case seems to be consistent with the metaplasia theory.

ULM had a variable term in the reported literature. It is needed to clarify the used terms and the differential diagnosis of ULMs. Adenomyoma is a benign lesion typically originating within the uterus. Both adenomyoma and ULMs are comprised of endometrial tissue and smooth muscle. However, ULM has unique organ-like differentiation structure which composed of a central cavity lined by endometrium with surrounding smooth muscle tissue; but the endometrioid tissue in adenomyoma was randomly distributed in multiple foci; there is no organoid differentiation structure. ULM can be distinguished from adenomyomas by such traits. ULMs can also be differentiated from endometriosis by their characteristic uterus-like morphology and distinct smooth muscle component. ULM should also be differentiated from rudimentary uterine horn. Rudimentary uterine horn caused by developmental defects the middle and lower part of the unilateral Müllerian tube, about two-thirds accompanying uterus unicornis. It is often communicated with the well-developed uterus or connected with the exterior surfaces via fibrous tissue, and often linked with normal fallopian tube and ovaries. ULM is a solitary mass outside the uterus and is not associated with the uterus or fallopian tubes. In addition, the uterus is often free of deformity.

In conclusion, we have reported a very rare case of ULM in the broad ligament with acute abdominal pain. In the case presented herein suggested that the mass is most likely a benign mass formed by the endometrial stromal cells or pluripotent mesenchymal cells of the 2nd Müllerian system. In addition, a uterine myoma was located in the posterior wall of the uterus. The occurrence of uterine myoma was also related to hormone stimulation. Therefore, the present case seems to be consistent with the metaplasia theory. Its pathogenesis remains to be further studied.

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