



A case of uterus-like mass of the ovary associated with endometriosis



Yee Jeong Kim^a, Joeng Hae Kie^a, Jae Eun Chung^{b,*}

^a Department of Pathology, National Health Insurance Service Ilsan Hospital, South Korea

^b Department of Obstetrics and Gynecology, National Health Insurance Service Ilsan Hospital, South Korea

1. Introduction

Endometriosis is defined as the presence of uterine endometrial glands and stroma in ectopic locations accompanied by variable degrees of smooth muscle metaplasia. A uterus-like mass (ULM) is presented with an endometrial cavity composed of well-differentiated endometrial glands and stroma surrounded by well-organized smooth muscle bundles, showing a strikingly structural similarity with that of a normal uterus. What triggered the ULM to acquire such a structural completeness and whether it should be considered as a rare form of endometriosis remains to be explored.

2. Case report

A 35-year-old nulliparous woman visited the emergency room due to excruciating dysmenorrhea that did not subside with the usage of conventional pain killers. She had no past history of sexual intercourse and she had never visited a gynecologist before. Her menarche started when she was 14-years-old. The dysmenorrhea was evident in her early 20's and it aggravated even with incremental adjustment of the pain killer dosage. As she had no previous sexual intercourse, transabdominal ultrasonography was taken which showed bilateral ovarian cysts, each measuring 6 cm and 7 cm, respectively. Her CA 125 level was elevated to 112 U/mL. Under the tentative diagnosis of bilateral ovarian endometriomas, exploratory laparotomy was planned. A right ovarian cystectomy was done with removal of the endometriotic cyst while leaving normal ovarian tissue to save her fertility. Her left ovary was enlarged to 7 cm. Left ovarian cystectomy could not be done due to the unusually hard consistency of the mass. Endometriotic spots were scattered throughout the pelvis and adhesion bands were noted. The pathologic diagnosis of the right ovarian cyst confirmed an endometrioma, as expected. The cut surface of the left ovarian mass showed a central endometrial cavity surrounded by bundles of smooth muscle cells; the general architecture resembled a normal uterus (Fig. 1). Histologically, the central cavity was lined with tubular glands and cylindrical epithelium surrounded by stroma. This central endometrial cavity-like structure was again surrounded by thick bundles of smooth muscle cells (Fig. 2). Immunostaining with desmin was

positive, as is noted in well-differentiated smooth muscle cells (Fig. 3). The left ovarian mass was pathologically confirmed as a ULM. To rule out other accompanying congenital anomalies, a computed tomography scan was taken after pathologic confirmation. No congenital anomaly of the urinary or skeletal system was found.

3. Discussion

About 30 cases of a uterus-like mass (ULM) arising in sites other than the uterus itself have been reported since Cozzutto's first observation in 1981 [1–12]. As the nomenclature implies, ULM presents with a well-differentiated endometrial cavity surrounded with smooth muscle bundles, showing a strikingly structural similarity with that of a normal uterus.

Endometriosis is defined as the presence of uterine endometrial glands and stroma in ectopic locations accompanied by a variable degree of smooth muscle metaplasia. It was first described by Sampson as scattered peritoneal implants of regurgitated endometrial tissue through fallopian tubes that caused dysmenorrhea [13]. Although widely accepted as the pathophysiologic mechanism of endometriosis, Sampson's retrograde menstruation theory cannot explain endometriosis developing in males and females without a uterus, therefore not experiencing menstruation [6]. Other theories regarding the pathogenesis of endometriosis include metaplasia of the coelomic epithelium, induction of the Mullerian rests also known as mullerianosis, lymphatic or hematogenous spread, adult stem cell activation via bone marrow or endometrium, genetic vulnerability and activation of endocrine disrupting chemicals [14–16].

Embryologically, the coelomic cavity is formed from the mesoderm and develops into pericardial, pleural and peritoneal cavities lined by mesothelium (coelomic epithelium). A part of this coelomic epithelium lying lateral to the urogenital ridge invaginates and coalesces to form the Mullerian duct. The Mullerian duct gives rise to the fallopian tubes, uterus, cervix and upper vagina [17]. As the coelomic epithelium plays the role of a common ancestor to the epithelium overlying the peritoneum and the Mullerian duct, Lauchlan named the peritoneum the secondary Mullerian system to acknowledge its potential to differentiate into Mullerian-directed epithelium and stroma. The secondary

* Corresponding author at: Department of Obstetrics and Gynecology, National Health Insurance Service Ilsan Hospital, 100, Ilsan-ro, Ilsan-donggu, Goyang-si, Gyeonggi-do 410-719, South Korea.

E-mail address: jiupark@naver.com (J.E. Chung).

<http://dx.doi.org/10.1016/j.crwh.2017.08.001>

Received 26 July 2017; Accepted 31 August 2017

Available online 08 September 2017

2214-9112/ © 2017 The Authors. Published by Elsevier B.V. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

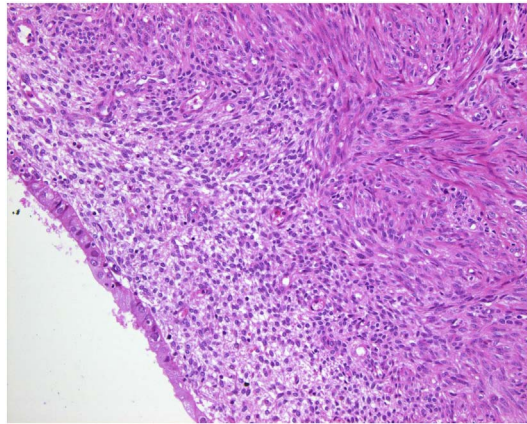


Fig. 1. Gross presentation of the uterus-like mass.



Fig. 2. The central cavity lined with endometrial epithelium and stroma is surrounded by thick bundles of smooth muscle ($\times 100$, H-E stain).

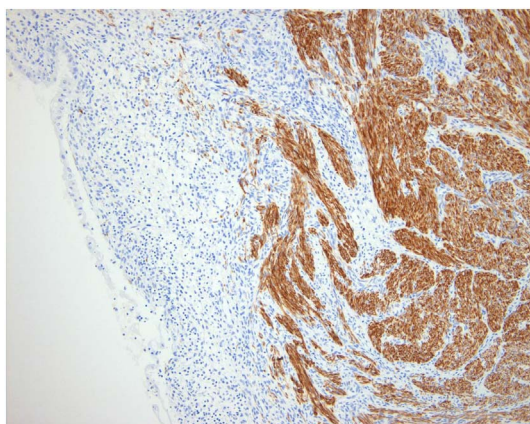


Fig. 3. Immunostaining with desmin is positive in the smooth muscle cells ($\times 100$, desmin).

Mullerian system encompasses Mullerian-type epithelium located outside the cavities of the original Mullerian duct such as the ovarian surface, pelvic peritoneum, omentum and retroperitoneal lymph nodes [18]. The epithelium covering the ovary is derived from coelomic epithelium. Invagination of the ovarian epithelium into the ovarian parenchyma results in an inclusion cyst. The epithelial lining of this inclusion cyst acquires the characteristics of Mullerian duct epithelium through coelomic metaplasia. The three most common subtypes of ovarian epithelial neoplasm are serous, endometrioid, and mucinous types. Morphologically, cell types of the above mentioned ovarian epithelial neoplasm resemble that of the fallopian tube, endometrium and endocervix, respectively. Therefore it can be postulated that coelomic metaplasia has resulted in the transformation of the invaginated normal ovarian epithelium into an ectopic endometrial lining and ovarian stromal cells into smooth muscle metaplasias forming an endometrioma of the ovary [19].

In this case, the patient had a right ovarian endometrioma and left ovarian ULM. These two seemingly different structures might share coelomic metaplasia as the histogenetic mechanism: coelomic metaplasia resulting in endometrial glands and stroma with smooth muscle metaplasia forming the right ovarian endometrioma and further organization of each component into structurally intact ULM in the left ovary. ULM might be considered a rare manifestation in the spectrum of endometriosis development. Of the 30 cases reported so far, 25 (83%) revealed variable manifestations of endometriosis such as concomitant ovarian endometriomas, accompanying peritoneal endometriotic implants with adhesion bands, a history of previous endometriosis surgery or endometrioid and clear cell ovarian carcinoma development (Table 1) [1–3,5,6,8–10,12]. ULM cases without concomitant endometriosis typically appeared outside the pelvic cavity, therefore suggesting heterotopia or choriostoma formation as the underlying mechanism [4,7,11].

ULM, under the postulated mechanism of coelomic metaplasia, can be considered a rare event in the spectrum of endometriosis development. The inducing factor for the coelomic metaplasia as well as the intense metaplastic transformation resulting in the structural integrity of a miniature uterus remains to be explained, as this might shed light on the complex and enigmatic pathogenesis of endometriosis.

Table 1
Reported cases of uterus-like mass.

Authors (published year)	Age	Location	Associated conditions	Histogenesis
Cozzutto (1981) [1]	31	ovary	Unilateral renal agenesis ULM not connected to the uterus No residual ovarian parenchyma attached to ULM Total replacement of the ovary by ULM?	Anomaly? Metaplasia?
Pueblitz-Peredo (1985) [2]	18	ovary	Double ureter, bifid pelvis Bilobated: endometrioma + ULM Size of ULM too large for a rudimentary horn Uterus bicornis unicollis-1 rudimentary horn?	metaplasia
Rahilly (1991) [3]	38	ovary	Bilated: ULM + endometrioid adenocarcinoma endometriosis	metaplasia
Horie (2000) [4]	59	Small bowel mesentery	Ectopic müllerian tissue	heterotopia
Redman (2005) [5]	50	Previous surgery site	History of TAH + BSO due to endometriosis Usage of estrogen patch	metaplasia
Shutter (2005) [6]	11	ovary	Pelvic kidney Residual normal ovarian parenchyma on the periphery of the mass No connection between ULM and the uterus	metaplasia
Sharma (2007) [7]	33	Conus medullaris	Ectopic müllerian tissue	Heterotopia
Kaufman (2008) [8]	24	Conus medullaris		
	57	Retroperitoneum	History of TAH + BSO due to endometriosis	Metaplasia
		History of	Usage of estrogen patch	
Liang (2010) [9]	17	Broad ligament	No müllerian anomaly Endometriotic spots	Metaplasia
Seki (2011) [10]	49	Inguinal subserosa	History of endometriotic cyst operation Accompanying endometriotic lesions	Metaplasia
Shin (2011) [11]	31	Sigmoid mesocolon	Ectopic müllerian tissue	Heterotopia
Nakakita (2014) [12]	67	ovary	ULM + clear cell carcinoma 14 years of follow up Natural history of ULM?	metaplasia

References

- [1] C. Cozzutto, Uterus-like mass replacing ovary: report of a new entity, *Arch. Pathol. Lab. Med.* 105 (1981) 508–511.
- [2] S. Pueblitz-Peredo, E. Luevano-Flores, R. Rincon-Taracena, F.J. Ochoa-Carrillo, Uterus-like mass of the ovary: endometriosis or congenital malformation? A case with a discussion of histogenesis, *Arch. Pathol. Lab. Med.* 109 (1985) 361–364.
- [3] M.A. Rahilly, A. al-Nafussi, Uterus-like mass of the ovary associated with endometrioid carcinoma, *Histopathology* 18 (1991) 549–551.
- [4] Y. Horie, M. Kato, Uterus-like mass of the small bowel mesentery, *Pathol. Int.* 50 (2000) 76–80.
- [5] R. Redman, E.J. Wilkinson, N.A. Masoll, Uterine-like mass with features of an extrauterine adenomyoma presenting 22 years after total abdominal hysterectomy and bilateral salpingo-oophorectomy: a case report and review of the literature, *Arch. Pathol. Lab. Med.* 129 (2005) 1041–1043.
- [6] J. Shutter, Uterus-like ovarian mass presenting near menarche, *Int. J. Gynecol. Pathol.* 24 (4) (2005) 382.
- [7] M.C. Sharma, C. Sarkar, D. Jain, V. Suri, A. Gaarg, S. Vaishya, Uterus-like mass of müllerian origin in the lumbosacral region causing cord tethering. Report of two cases, *J. Neurosurg. Spine* 6 (2007) 73–76.
- [8] Y. Kaufman, A. Lam, The pelvic uterus-like mass: a primary or secondary müllerian system anomaly? *J. Minim. Invasive Gynecol.* 15 (2008) 494–497.
- [9] Y.J. Liang, Q. Hao, Wu YZ, B. Wu, Uterus-like mass in the left broad ligament misdiagnosed as a malformation of the uterus: a case report of a rare condition and review of the literature, *Fertil. Steril.* 93 (2010) e13–6.
- [10] A. Seki, A. Maeshima, H. Nakagawa, J. Shiraiishi, A subserosal uterus-like mass presenting after a sliding hernia of the ovary and endometriosis: a rare entity with a discussion of the histogenesis, *Fertil. Steril.* 95 (2011) e15–9.
- [11] S.Y. Shin, H.J. Kim, Y.W. Kim, K.Y. Lee, CT characteristics of a uterus-like mass in the sigmoid mesocolon, *Br. J. Radiol.* 84 (2011) e1–3.
- [12] B. Nakakita, K. Abiko, Y. Mikami, A. Kido, Clear cell carcinoma arising from a uterus-like mass, *Pathol. Int.* 64 (2014) 576–580.
- [13] J.A. Sampson, Peritoneal endometriosis due to menstrual dissemination of endometrial tissue into the peritoneal cavity, *Am. J. Obstet. Gynecol.* 14 (1927) 422–469.
- [14] M.M. Melicow, M.R. Pachter, Endometrial carcinoma of the prostatic utricle (uterus masculinus), *Cancer* 20 (1967) 1715–1722.
- [15] D.L. Rosenfeld, B.D. Lechner, Endometriosis in a patient with Rokitansky-Kuster-Hauser syndrome, *Am. J. Obstet. Gynecol.* 139 (1981) 105–107.
- [16] R. Burney, L. Giudice, Pathogenesis and pathophysiology of endometriosis, *Fertil. Steril.* 98 (2012) 511–519.
- [17] T. Parmlay, Embryology of the female genital tract, Balustein's pathology of the female genital tract, 3rd ed., Springer-Verlag, New York, 1987, pp. 1–14.
- [18] S.C. Lauchlan, The secondary müllerian system, *Obstet. Gynecol. Surv.* 27 (1972) 133–146.
- [19] M. Nisolle, J. Donnez, Peritoneal endometriosis, ovarian endometriosis, and adenomyotic nodules of the rectovaginal septum are three different entities, *Fertil. Steril.* 68 (1997) 585–596.