Case report

Asymptomatic uterus-like mass (ULM) of the extraperitoneal space – a case report of a rare finding with unusual clinical presentation and review of the literature

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The paper presents a case of a uterus-like mass (ULM), a rare type of tumour of the female reproductive system, which did not present any clinical symptoms described in other cases of ULMs. There are 35 reported cases of this type of tumour. It is defined as a lesion composed of smooth muscle-like stromal cells with a central cavity lined with endometrial type epithelium. There are three theories on the pathogenesis of ULMs which we discuss along with clinical presentation, diagnostic features, treatment options and potential oncological implications of this type of tumour, based on our case, and the review of the literature.

Key words: ULM, uterus-like mass, endomyometriosis, pelvic tumour.

Introduction

Uterus-like mass (ULM) is a rare type of tumour composed of smooth muscle cells with a central cavity lined with endometrium. It was first described in 1981 by Cozzutto, who found features resembling a uterus in an ovarian lesion [1]. Until now there are 35 cases of ULM reported in English-language literature [2, 3, 4, 5]. The typical clinical manifestation includes: recurrent pelvic pain, dysmenorrhea, intermenstrual bleeding and occasionally dysuria. Elevated serum CA125 is also a common finding [3, 6, 7, 8]. There are reported cases of endometrial [9] and clear-cell [4] carcinoma arising from a ULM. Reported location sites of ULMs include: the ovary [1, 9, 10], the uterine cervix [11], the broad ligament [12, 13], the uterus [14, 15], the small bowel mesentery [16], the colon mucosa [2], the ovarian ligament [17] and scrotum [5]. We present a case of a 15 cm ULM arising in the extra-peritoneal space of the pelvis, ranging from the cavum Retzii medially, to the right inguinal canal and right obturator fossa laterally, and to the bifurcation of the right common iliac vessels cranially. Preoperative imaging raised suspicion of ovarian malignancy, while the patient was otherwise asymptomatic. Afterwards, potential pathogenesis of ULMs is discussed, along with a review of the literature.

Case report

Our patient is a 32-year-old nulliparous female referred to an Oncology Centre by a GP, because of a pelvic mass which presented features suspicious of

ovarian malignancy, found during a routine abdominal ultrasound scan. The interview revealed no previous history of gynaecological or other diseases, no history of malignancy in the family, and was also negative for allergies and addictions. The patient did not present any subjective symptoms. As she was a virgin, the first gynaecological examination was performed per rectum. It revealed an indolent, partly solid, partly cystic mass at the right side of the pelvis. The largest palpable dimension was about 6 cm and the walls of the tumour were irregular. Surgical abdominal examination pointed out the same tumour, and no other abdominal symptoms. The laboratory tests showed normal levels of CA 125; CEA; CA 19.9. Other tests such as hematologic morphology, creatinine, eGFR, INR, APTT, CRP, TSH, fasting glucose level, protein levels, and the examination of the urine were all in the normal range. The CT scan of the pelvis and abdomen had shown a tumour in the place of the right ovary – the dimensions of the solid part of the lesion were 90 × 65 mm and the density was uneven, while the cystic part had a dimension of 123 mm, and grew downwards and to the front displacing the urinary bladder. The presentation of the kidneys was normal, and no signs of dilation of the ureters were found. The gallbladder, pancreas, adrenal glands and lymph nodes were normal. In the 3rd segment of the liver a 43 mm lesion amplifying after administration of contrast, with a central hypodense zone, was found and referred to further inspection with MRI, which revealed no signs of suspected malignancy. The hepatic lesion was left under observation, without the need for H-P examination.

After a multidisciplinary consultation of the patient, the team decided to treat the patient surgically. The patient was willing to retain fertility, and agreed to undergo only unilateral adnexectomy or preferably tumorectomy no matter of the intraoperative histological examination result. As the tumour appeared too big for laparoscopic excision, a Pfannenstiel suprapubic laparotomy had been chosen as a minimally invasive technique for this case. The cystic part of the lesion has found to be filling the cavum Retzii and continued laterally towards the right iliac vessels. It was extracted using blunt dissection, without perforation of the capsule. The solid part spread to the entrance of the external iliac vessels to the inguinal canal laterally, posteriorly it reached the obturator fossa and cranially ended at the bifurcation of the common iliac vessels. As its macroscopic appearance somewhat resembled a kidney and the distal part of the ureter was involved in the lesion, an intraoperative ultrasound had been performed before excision, and shown an isoechoic mass with a central cavity and a small amount of hypoechoic fluid within the cavity. A normal right kidney had also been found by ultrasound in its typical location. The ureter had been freed from the mass and the tumour excised and sent for an intraoperative H-P examination, which revealed no symptoms of malignancy. Inspection of the abdominal cavity had shown normal uterus and adnexa. No other macroscopic abnormalities had been found (Figs. 1, 2).

The postoperative course was uneventful and the patient had been released in the 3rd day after surgery.

The final histopathology report had shown a polycyclic tumour comprised of a 14 cm cystic part and a 11.5 cm solid part. The total size of the lesion was 15,5 × 14 × 9 cm. The inner surface of the cystic part was smooth and the fluid serous and clear. The solid part had a thick fibrous wall and a central cavity measuring 6,5 cm. Microscopically the cavity was lined with endometrial type epithelium surrounded by fusiform stromal cells with features of smooth muscle tissue with thick walled blood vessels. The IHC staining revealed positive SMA (smooth muscle actin) and desmin reactions as well as came out positive for ERs (oestrogen receptors) and PgRs (progesterone receptors) within the stromal cells, which corresponds to uterine smooth muscle tissue. The epithelial cells were positive for AE1/AE3 CK (cytokeratin) and CK7 which is characteristic to epithelial cells of the reproductive system eg. endometrial. Positive nuclear PAX-8 staining within the epithelium may suggest the Müllerian system as the origin of the tumour. Surprisingly the epithelium of what negative to ERs and PgRs. The tumour was consistent with the presentation of a ULM (Table I, Fig. 3). 6 months of follow-up showed no signs of recurrence nor any complications.

Table I. IHC results

Antibody	Reactivity	
	Glandular cells (epithelium)	Stromal cells (muscle)
CK AE1/AE3	+	–
CK7	+	–
PAX 8	+	–
ER	–	+
PgR	–	+
SMA	–	+
Desmin	–	+
CA-125	–	–

Discussion

The ULM is an unusual gynaecological finding defined as a smooth muscle tumour with a central cavity, that is lined with endometrium [13, 18]. In the review of the English-language literature to date, we found

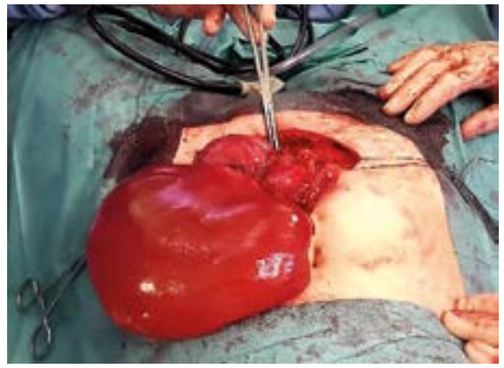

Fig. 1. Intraoperative presentation of the cystic part

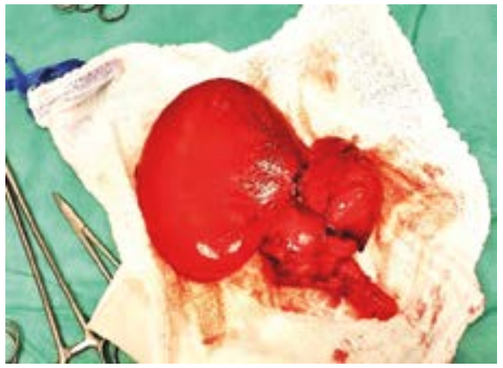

Fig. 2. Postoperative presentation of the tumour

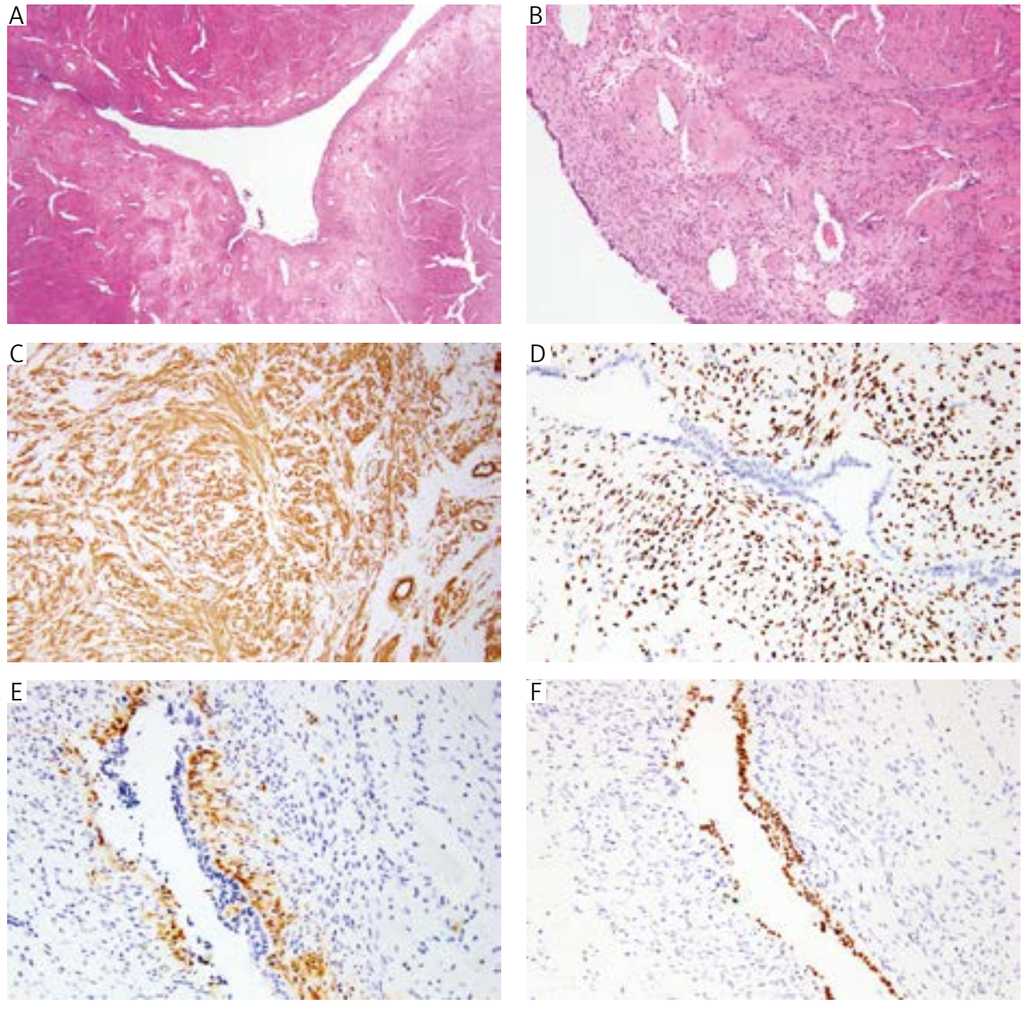

Fig. 3. HP presentation of the lesion

only 35 described cases of the phenomenon [2, 3, 4, 5]. ULMs localize themselves in various parts of the female reproductive system, the pelvic wall, uterine and ovarian ligaments, the bowel mucosa and the bowel mesentery [1, 2, 3, 6, 7, 8, 9, 10, 11, 12, 13, 14, 15, 16, 17]. There is also a case of a ULM found in the scrotum of a 82-yo male who underwent hormonal therapy [5]. Common clinical symptoms are non-specific and include lower abdominal pain (LAP), lower back pain (LBP), vaginal spotting, dysmenorrhea, vulvar itching, constipation and dysuria [2]. The age of women diagnosed with ULM ranges from 12 to 59 years, while the man was 82 [1, 2, 3, 4, 5, 6, 7, 8, 9, 10, 11, 12, 13, 14, 15, 16, 17]. Different sizes of ULMs range from 2,5 to 21 cm [2]. Imaging techniques used in the diagnostic process include USG, CT, and MRI. The image is most often described as a homogenous mass with a central cavity, and occasionally a cystic part on the outer side of the lesion [3, 6]. A more detailed description of the MRI can show a low-signal zone surrounding the endometrium, which corresponds to the junctional zone between the endometrium and the myometrium [6]. It seems that this zone could also be visualized by high-quality ultrasound, given good sonographic conditions.

Our case is the first reported clinically asymptomatic ULM localized within the pelvis, nevertheless considering the location and size of the lesion, as well as the involvement of the distal part of the ureter it would probably soon manifest with lower abdominal pain, lower back pain, or renal colic. The probable cause for the lack of common ULM symptoms in or patient such as dysmenorrhea, LAP or LBP is the lack of ERs and PgRs in the endometrium of the lesion. We think that this resulted in a ULM with an endometrium independent of the woman's menstrual cycle, and thus only a scarce amount of blood accumulated within the cavity of the tumor.

There are three main theories on the pathogenesis of ULM: 1. Müllerian duct fusion defect, 2. Heterotopia, 3. Metaplasia in the "secondary Müllerian system" [5, 7, 12, 13, 14, 16]. The Müllerian duct fusion defect theory, also called the congenital anomaly theory refers to the lack of fusion of the Müllerian ducts throughout their path, which is the common cause of uterine and vaginal anomalies. It was first proposed by Rosai in response to the original Cozzutto paper in 1982, suggesting that the lesion was an example of a uterus unicornis [1, 16, 19]. As in most cases of the ULM there was an anatomically correct uterus and fallopian tubes present, with no accompanying anomalies in the urinary system, this explanation seems highly unlikely. The heterotopia theory was presented by Peterson *et al.* while reporting a case of a uterus-like mass in the ileum in a 12-year-old with multiple anomalies, such as sacral agenesis, sacrococcygeal teratoma and defects of the lower genital and intestinal tracts. The authors indicated that neither the anomaly nor the metaplasia theory provided sufficient explanation for the presence of a ULM in the small intestine [13, 16, 20]. The metaplasia theory is most commonly used to explain the presence of ULM. It is based on the concept of a "secondary Müllerian system" which was proposed by Lauchlan in 1972, and assumes that the peritoneal mesothelium and the adjacent connective tissue is embryologicaly closely related to the Müllerian system and retains the potential to differentiate into Müllerian structures such as fallopian tubes, the uterus or vagina [12, 16, 21]. This theory also is the best explanation of our case of ULM arising in the connective tissue of the pelvic extra-peritoneal space.

Uterus-like masses also have an oncological inclination. Pai *et al*. observed elevated levels of serum CA-125 in three patients with ULM and found a coincidence with breast cancer. His observations led to a conclusion that ULMs are hormone dependent and he had classified them as a form of endometriosis [7, 10]. There are also cases of an endometrioid adenocarcinoma reported by Rahilly *et al*. and a clear-cell carcinoma reported by Nakakita *et al.,* both arising from a ULM [4, 9]. As those malignancies, both fit into the pathway of the carcinogenesis of endometriosis-related ovarian neoplasms, the more likely and important the association of ULM with endometriosis seems [4].

Conclusions

We have encountered a rare case of a ULM. As the preoperative presentation of the lesion resembles ovarian malignancies, thorough biochemical analysis and diagnostic imaging has been performed before treatment. Our patient did not present any subjective symptoms, which was probably caused by the lack of PgRs and ERs in the epithelium of the presented ULM. The surgery required very careful preparation of the tumor because of the proximity of the external and internal iliac vessels, and the involvement of the ureter. The postoperative course was uneventful, and the patient remains in follow-up.

Though rare ULMs should be considered when diagnosing pelvic masses. After review of the literature we think that radical excision is the best treatment option as there are reported cases of malignancies within this type of tumors. Minimally invasive procedures are advised where applicable.

The authors declare no conflict of interest.

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