Unicornuate uterus associated with retrosigmoid ovary and unilateral renal agenesis: a case report and a review of the literature

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ABSTRACT

Unicornuate uterus is a malformation characterized by the unilateral development of a single Müllerian duct during embryogenesis, while the contralateral part is either incompletely developed or absent. It can be associated with a symptomatic presentation, namely chronic cyclic pain, and with adverse reproductive and obstetric outcomes. Transvaginal ultrasound examination, sometimes requiring 3D imaging, is used to diagnose and classify this condition; further investigation may include magnetic resonance imaging (MRI). In this report, we present the case of a healthy 22-year-old woman reporting irregular menstrual cycles who underwent transvaginal ultrasound examination at our obstetrics and gynecology unit. Ultrasonography revealed a unicornuate uterus with absence of rudimentary horn and ipsilateral adnexa in the pelvis. MRI was requested, and showed the presence of a normal ovary located in an ectopic retrosigmoid location, and ipsilateral renal agenesis. These abnormalities may quite frequently occur in the presence of uterine malformations such as unicornuate uterus. A correct classification of such defects and the associated abnormalities is needed in order to be able to counsel patients about potential issues regarding fertility and preservation of renal function, and to discuss the possibility of medical or surgical management of symptoms or associated complications. Ultrasound examination could miss associated defects, and an MRI scan is often necessary to define the diagnosis.

KEYWORDS

Uterus; ovary, congenital abnormalities, solitary kidney, ultrasonography, magnetic resonance imaging.

Introduction

Congenital malformations of the female genital tract can result from embryological maldevelopment of the Müllerian or paramesonephric ducts. A unicornuate uterus, or hemi-uterus, is defined as a malformation in which the uterus is formed by the unilateral development of a single Müllerian duct during embryogenesis, while the contralateral part is either incompletely developed or absent [1]. The single uterine horn is usually connected to a normal salpinx and cervix; the failed Müllerian duct, on the contrary, may undergo transformation into a rudimentary structure showing various degrees of development, which may or may not communicate with the functioning horn [1, 2]. This class of uterine malformation has a very low prevalence, being estimated to occur in about 1 out of 1000 women in the general population [3]; instead, it is found in 3–13% of women with Müllerian defects [2, 3].

The clinical significance of the unicornuate uterus is due to the fact that it carries an increased risk of miscarriage and obstetric complications; there is also the possibility of symptoms due to the cyclical menstrual modifications occurring in the rudimentary horn. A unicornuate uterus is associated with a significant risk of ectopic pregnancy, first and second trimester abortion, preterm labor, malpresentation at delivery, low birth weight, perinatal mortality and intrauterine fetal demise, with a reduced live birth rate [3-5]; the prevalence of this defect is therefore higher (about 5 in 1000 women) in high-risk populations, including women with infertility or recurrent miscarriage. A functional rudimentary horn contains endometrium which undergoes changes with every menstrual cycle; endometrial shedding in obstructed non-communicating horns typically results in chronic cyclic pain. In unicornuate uteri with communicating horns, there is the possibility of pregnancy developing inside the rudimentary cavity, a situation associated with a higher risk of uterine rupture and placental attachment abnormalities [6]. Rare cases of tumors arising in the rudimentary horn are also described in the literature [7].

The diagnosis is usually made on the basis of pelvic ultrasound examination showing an abnormal uterine shape. A recent consensus on the classification of congenital abnormalities of the female genital tract included unicornuate uterus,

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defined as "hemi-uterus", in class U4, which is further divided into two sub-classes, i.e., with or without a functional rudimentary cavity [1]. A traditional 2D ultrasound scan generally allows detection of this abnormality, which can be further explored by second-level examinations such as 3D ultrasonography or magnetic resonance imaging (MRI), in order to better define the uterine hemi-cavity. Ultrasound examination can show the presence of a rudimentary horn, with a wide variety of appearances according to the degree of development. Moreover, it can help to raise suspicion of associated congenital abnormalities of the ovary or urinary system.

Case report

A 22-year-old woman with irregular menstrual cycles was referred to the Obstetrics and Gynecology Unit at the Mauriziano Umberto I Hospital in Turin to undergo a pelvic ultrasound examination. The patient was in good health and did not complain of any other gynecological symptom; in the past years, she had experienced some episodes of low back pain on the left side, which were diagnosed as renal colic and managed conservatively with painkillers. The patient had never undergone an ultrasound examination before, either vaginal or abdominal.

Pelvic ultrasound examination was performed with an Affiniti 70 ultrasound machine equipped with a C9-4v endocavitary probe with a 4.0–9.0 MHz frequency range (Philips, Amsterdam, the Netherlands, 2013). The scan revealed the presence of a unicornuate uterus, with a typical appearance on 2D examination. The uterus, due to the presence of the single horn, was deviated laterally to the right side of the pelvis (Figure 1); when scanning in transverse planes from the cervix to the fundus, the endometrium appeared tapering and increasingly displaced on the right side, in contrast to a normally appearing endometrium, which is usually characterized by a progressive symmetrical widening of the endometrial stripe. No rudimentary horn could be seen in the pelvis on the contralateral side. Examination of the adnexa showed, on the right side, an apparently normal ovary, located posteriorly to the uterine fundus, and no signs of tubal anomalies. On the contrary, on the left side, no ovary or salpinx could be seen, either in their normal location or in other sites of the pelvis; an ill-defined hypoechoic area of uncertain nature was seen posterior to the distal part of the sigmoid colon. Finally, combined transvaginal and transabdominal scan showed an apparently normal kidney on the right side, without any sign of dilation of the urinary tract, whereas no kidney could be seen on the left side (Figure 2).

In order to better define these abnormalities, an MRI examination was requested; the patient was therefore referred to the Radiodiagnostics Unit at our hospital. MRI confirmed the suspicion of unicornuate uterus, with normal cervix and vagina and normal appearance of the right adnexa. On the left side, the scan showed an image that seemed to correspond to an ectopic ovary located dorsally and laterally to the sigmoid colon (Figure 3).

Figure 1 Transvaginal pelvic ultrasound scan showing the midsagittal plane of the uterus, which is deviated laterally to the right side of the pelvis; the right ovary can be observed, located posterior to the uterine fundus.

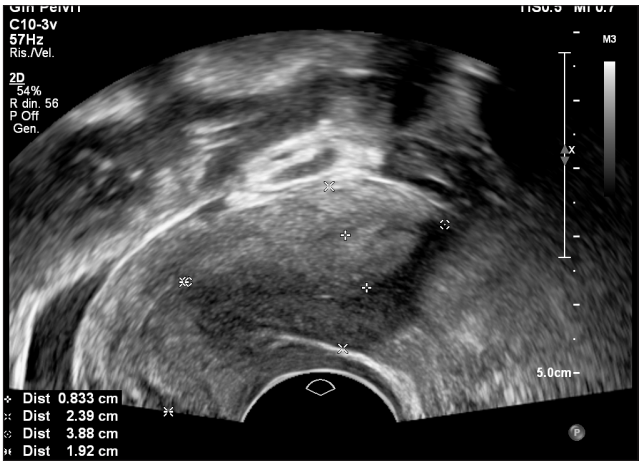


Figure 2 Transabdominal ultrasound scan of the left upper quadrant in the sagittal plane, showing the spleen; the left kidney is not visualized in its normal location.

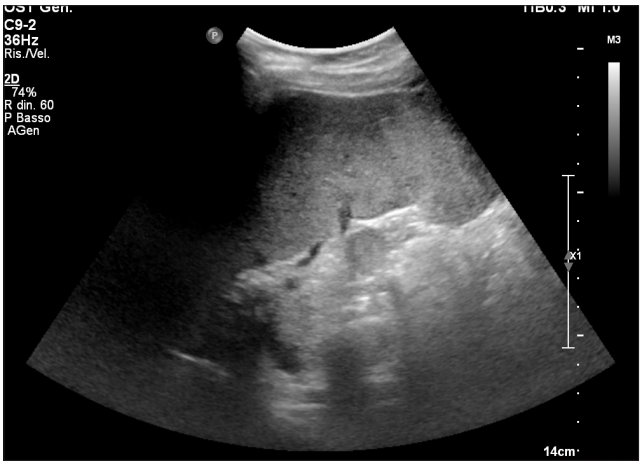
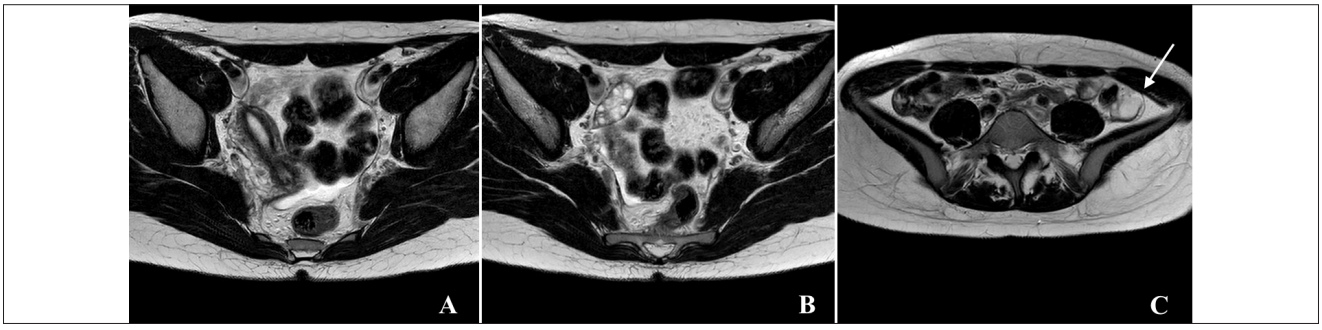


Figure 3 Magnetic resonance imaging scan with T2-weighted axial images showing the unicornuate uterus (A) and the normal ovary (B) on the right side of the pelvis; in the cranial plane, the ectopic left ovary (arrow) can be seen, positioned dorsally and laterally to the sigmoid colon (C).



The ovary was characterized by microcysts and enlarged size, without any apparent lesion. MRI also detected a structure attached to the ectopic ovary, which could be interpreted as a rudimentary horn with no functional endometrial cavity, and also confirmed the suspected agenesis of the left kidney; on the right side, no urinary tract abnormalities were identified, except for a slightly increased size of the right kidney, which could be due to compensatory hypertrophy.

Our patient did not present a significant burden of symptoms, except for infrequent low back pain on the left side; this was most probably related to the sporadic rupture of corpus luteum cysts developing in the ovary located in the retroperitoneal space, since no functional endometrium was seen in the rudimentary horn.

Counseling was carried out to explain the different possibilities with regard to the management of this condition; given the patient's young age it was decided to opt for follow up, abstaining from surgical removal of the ovary in order to prevent possible loss of ovarian function. She was also referred for a urological evaluation. The patient was not interested in offspring for the time being, but she was informed about fertility issues associated with her malformation and the possibility of early referral to a fertility center.

Discussion

The present report concerns a case of unicornuate uterus, classifiable as a U4b malformation according to the classification proposed by the European Society of Human Reproduction and Embryology (ESHRE) and the European Society for Gynecological Endoscopy (ESGE) [1]. In this patient, the condition was associated with ovarian malposition (an ectopic ovary was located in the retrosigmoid region) and also with congenital unilateral renal agenesis, affecting the side on which the Müllerian duct had failed to develop.

Ovarian malposition has a higher incidence in women with congenital uterine anomalies, and its prevalence is significantly increased in the presence of defects, such as unicornuate uterus, in which the uterus is absent or only partially present [8, 9]. The ectopic ovary may be attached to the uterus, colon, bladder or kidney, or located in the omentum, mesentery or inguinal canal. This condition usually derives from a congenital defect in the normal development of the pelvic organs, while some cases are suspected to be the result of auto-amputation of the ovary (e.g., because of torsion) with subsequent auto-transplantation in other sites, mostly the Douglas pouch [10-12]. In 1991, Lachman and Berman proposed a classification of ectopic ovary, dividing the condition into three categories: post-surgical implant, post-inflammatory implant, and true embryologic [13]. The ovaries initially develop in a para-renal location, before descending into the pelvis, guided by the gubernaculum, during the third month of fetal life [14]. True ectopy is thought to be due to incomplete caudal descent of the ovary into the pelvis or to delayed development of the portion of the urogenital ridge that gives rise to both the gonad and the Fallopian tube. This results in attachment of the upper ovarian pole to an area above the level of the common iliac vessels [2].

Although ovarian maldescent also occurs in patients with a normal uterus, its incidence is increased in women with Müllerian duct anomalies, with a higher association in those with unicornuate uterus: its incidence is reported to be 20% when the uterus is absent (Rokitansky-Küstner-Hauser syndrome) and as high as 42% in cases of unicornuate uterus [2, 15].

The ectopic ovary may harbor the development of adnexal masses, whose detection can be greatly impeded by the malposition, or even impossible on a traditional pelvic ultrasound scan [16-19]. The ovary, notably in presence of masses, can have a clinical presentation consisting of sporadic or recurrent abdominal pain, or even acute abdomen in the case of events such as torsion or rupture of cysts [20].

Unicornuate uterus is also linked to a higher incidence of renal abnormalities, which are estimated to have a prevalence up to 40% in carriers of the malformation. Associated congenital abnormalities include ectopic kidney (i.e., in the pelvis), renal agenesis, double renal pelvis and horseshoe kidney [21-24]. Such associations have been further highlighted in women with ectopic ovary, which can be accompanied by maldevelopment of the genital and urinary tracts, notably renal agenesis, with a higher prevalence when both uterine and ovarian congenital defects are present [9, 25, 26].

An MRI scan is often necessary in the evaluation of patients with congenital abnormalities of the genital tract. This imaging study allows better definition of the shape of the uterine cavity, and it can help in the detection of rudimentary Müllerian structures — these can easily be missed on ultrasound examination — and in assessing the possible presence of endometrial activity within them. Second, MRI can locate an ectopic ovary, allowing its examination to rule out adnexal lesions and helping in differential diagnosis when the ovary mimics a gastrointestinal or retroperitoneal space lesion [27, 28]. Due to its superior soft tissue contrast and multiplanar imaging capabilities, MRI has been shown to be more sensitive than ultrasound in the detection of an undescended ovary. In some cases, ovarian stimulation, e.g. with clomiphene, is necessary to better visualize the ectopic ovary [2]. Finally, MRI is mandatory for a full examination of the urinary tract, conducted to look for abnormalities that can be associated with congenital uterine and ovarian malformations. Use of MRI is recommended in all patients with suspected complex anomalies or in diagnostic dilemmas [29]. The MRI examination can typically be done without contrast, but this decision can be left to the discretion of the radiologist. Because of the sensitivity of MRI, laparoscopy is seldom required to make the diagnosis, but may be appropriate in a patient presenting with pelvic pain [30].

A correct diagnosis, including a classification of the uterine malformation, is mandatory for counseling regarding fertility and the risk of pregnancy-related complications. The patient should be provided with information about the risk of impaired pregnancy outcomes, and the low evidence regarding reconstructive surgery as a means of improving them in cases of unicornuate uterus. In women with menstrual cycle-related symptoms, due to shedding of endometrium in a functioning non-communicating horn, imaging studies are important to identify the rudimentary structure and discuss the possibility of surgical removal.

Ectopic ovaries should be identified to allow monitoring for the development of neoplasms or benign functional lesions; a further specific case is the possibility of ovulation induction in women undergoing assisted reproduction techniques. Moreover, patients with symptoms related to this malformation (e.g., cyclical pain due to the rupture of corpus luteum cysts) might be referred for surgery. Given the low number of cases reported in the literature, no recommendations are available regarding the management of an asymptomatic ectopic ovary; however, conservative management appears to be a reasonable approach in order to avoid a reduction in ovarian reserve, since no data suggest a higher risk of developing neoplasms or acute complications such as torsion.

The detection of associated urinary tract abnormalities may require referral to a urology specialist and follow up to detect any change in renal function, which could become impaired over the years in patients with a single kidney.

Conclusions

The detection of a uterine malformation requires a thorough diagnosis including a correct classification of the defect and identification of any associated abnormalities involving the adnexa and the urinary system. An MRI scan is often necessary since ultrasound examination could miss associated defects.

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