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Case Report

Difficulties in the Diagnosis of Uterine Congenital Malformations: Ruling out a Unicornuate Uterus

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Abstract

Ruling out a unicornuate uterus might not be as simple as it seems. A wide differential diagnosis must be considered and for it, embryology for the correct understanding of the malformation has to be reminded, and several diagnostic tools to evaluate the female genitalia and urinary tract are needed.

INTRODUCTION

There is a varied amount of uterus congenital anomalies. Most of them are underdiagnosed due to alack of symptoms during childhood but start to show after menarche.Symptoms in the case of young women range from forms of dysmenorrhea, miscarriages or premature deliveries, but uterine anomalies might be also present in 2 to 3 percent of fertile women with normal reproductive outcomes [1]. So patients who are asymptomatic might realize they have a congenital anomaly after a routine exam is performed or through an ultrasound during pregnancy. In the study of these anomalies, hysteroscopy orhysterosalpingography (HSG) may be carried out to examine the uterus cavity, as well as 3D-ultrasound, magnetic resonance imaging (MRI) or a diagnostic laparoscopy to examine the external surface [2]. An intravenous pielography is also highly recommended to assess the urinary system.

CASE REPORT

A 28 year old, nulliparous asymptomatic woman, eumenorrheic and seeking pregnancy was referred to our hospital due to an ultrasound diagnosis of uterine malformation and adnexal cyst. On vaginal exam one cervix was seen but transvaginal ultrasound showed abicornuate uterus in which the connection between right and left horns was not visible (Figure 1), also a 4 cm paraovarian cyst on the left side was diagnosed. Renal ultrasound revealed both kidneys were orthotopically located.

HSG showed a right unicornuate uterus with normal tube (Figure 2A). Pielography demonstrated two normal kidneys with normal ureters (Figure 2B).The MRI reported two endometrial cavities without hematometra and left paracervical cyst (Figure 3A-B). Diagnostic hysteroscopy showed a single cervix with a normal cervical canalthat gave access to a right hemiuterus with an endometrial polyp and a visible ostium; from the endocervical canal it was not possible to access the left hemiuterus. Laparoscopy (Figure 3C) confirmed the suspicion: a unicornuate right uterus with a left cavitated non-communicating rudimentary horn, the left paraovarian cyst was subperitoneal and interpreted as a Gartner cyst.

The endometrial polyp and cyst were excised but since the patient was asymptomatic, no surgical procedure was performed on the uterus. The patient became pregnant afterwards and delivered a healthy baby at term through cesarean section due to breech presentation.

DISCUSSION

Unicornuate uterus (Class II of the American Fertility Society [3]-AFS-) with a rudimentary horn is not a common type of mullerian duct malformation which results from a normally developed müllerian duct fused with an underdeveloped contra-lateral one. Both ducts may connect through a fibro-muscular band or have no communication at all, being a loose band of tissue. So, the rudimentary horn may have a functional endometrial cavity communicated (Type IIa) or not (Type IIb), or be formed only by muscle with no endometrium (Type IIc). In all these situations, it is important to adequately identify the anomaly and to determine whether there is renal agenesis, since it could correspond to a more complex malformation [4].

Apart from the typical manifestations of unicornuate uterus (mainly premature labor or fetal mal presentation/breech

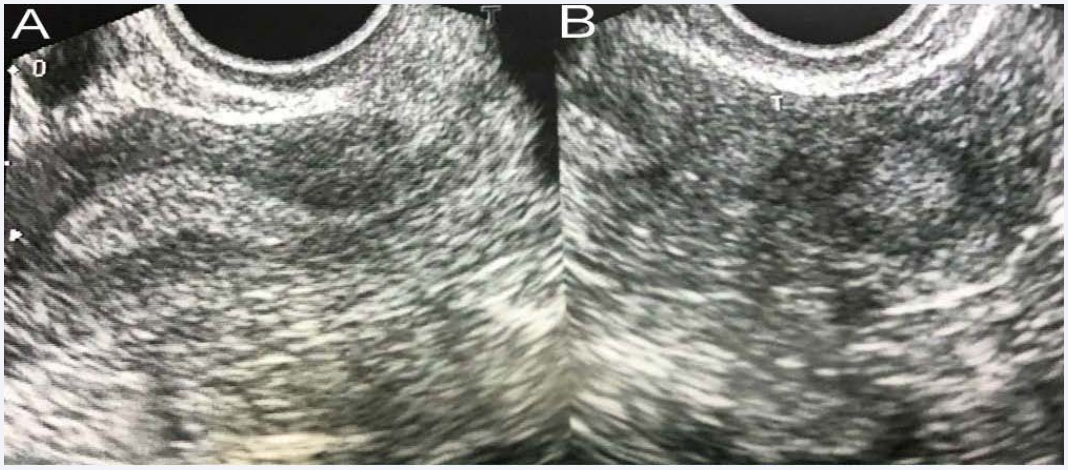

Figure 1 Transvaginal ultrasound showing a longitudinal and coronal section of a bicornuate uterus in which the endometrial connection between right and left horns is not visible.

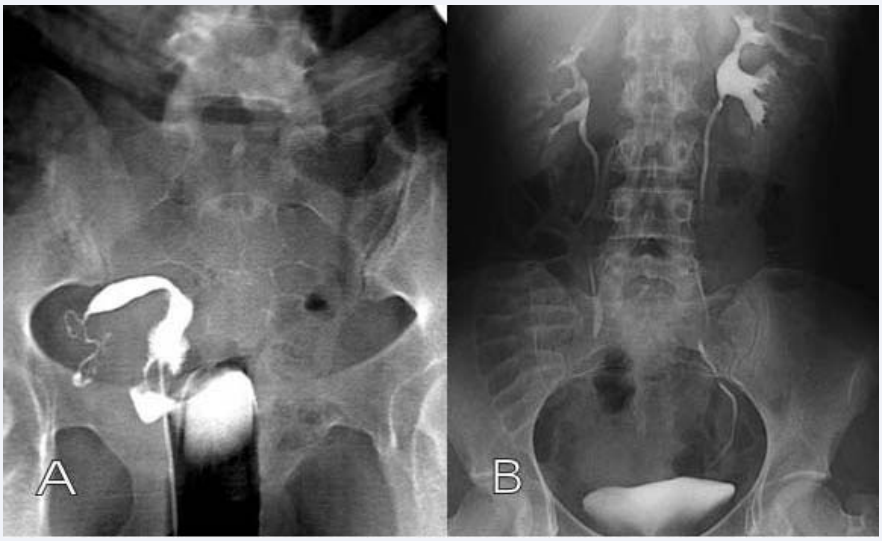

Figure 2 (A) Hysterosalpingography showing a right unicornuate uterus with normal tube (B) Intravenous pyelography showing normal kidneys and excretory system.

at birth [5]), in cases where a non-communicated functional endometrial cavity does exist (Type IIb of the AFS classification), complications may begin when the hormonal activity starts at menarche behaving as a complex malformation and providing serious clinical problems [4]. Patients may suffer from retrograde menstruation, presenting with dysmenorrhea, endometriosis and/or infertility [6], but sometimes there are no symptoms or adnexal pathology, as in our case. If there is no retrograde menstrual flow, hematometra may occur with a characteristically progressive increase of dysmenorrheal [7,8], neither present in our case. Other complications have also been described, as torsion of the rudimentary horn [9], or in the unlikely event that pregnancy occurs, rupture of the horn [10].

In such cases of unicornuate uterus with a cavitated non-communicated horn and both kidneys present, some differential diagnosis must be established. Firstly, a differential diagnosis with a cavitated accessory uterine mass (ACUM), which might be difficult. HSG and/or 3D-ultrasoundcan be done to differentiate them, but MRI is the preferred non-invasive modality [2,11]. ACUM is a non-communicating mass, usually located in the anterior wall of the uterus at the level of insertion of the round ligament. This anomaly is probably caused by duplication and persistence of ductal Mullerian tissue in a critical area at the attachment level of the round ligament, possibly related to a gubernaculum dysfunction [12]. Therefore, it needs to be classified separately as the uterine cavity is normaldue to a normal development of the müllerian ducts [13]. This entity characteristically presents at a young age, usually less than 30 years of age, with severe dysmenorrhea and chronic pelvic pain due to distention of the cavity caused by repeated bleeding [14].

Likewise, a segmentary atresia resulting in the Müllerian malformation has to be ruled out. In these situations, the external

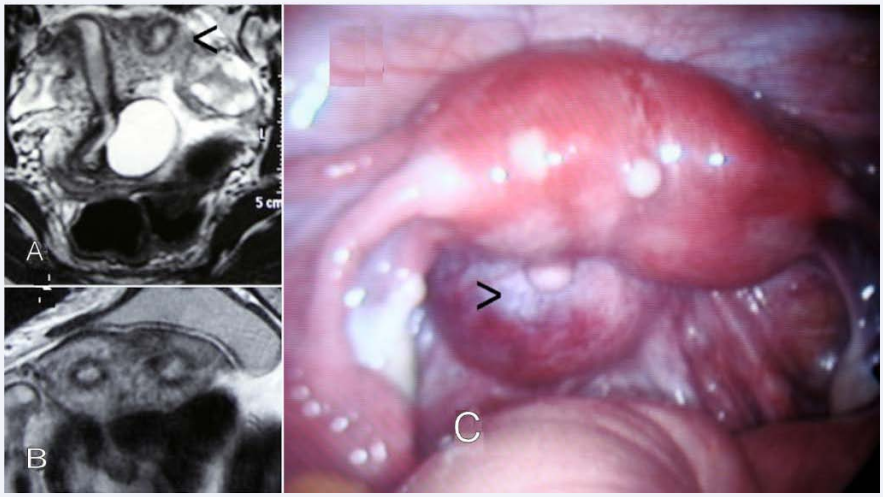

Figure 3 (A) Axial and (B) T2-weighted MR image (coronal cut)showing the left cavitated and rudimentary uterine horn (<).(C) Laparoscopic image of an apparently normal or very slightly arcuate uterus with normal adnexa, and a left retrocervical subperitoneal serous cyst (>) corresponding to a Müllerian remnant. (Modified from Acién and Acién, Hum Reprod Update 2016; 22: 48–69, Fig. 3B, with permission).

uterine morphology would be that of a bicornuate [15], or a unicornuate uterus with both uterosacral ligaments ending in its inferior portion and at isthmic portion a tract extending towards the thickened uterine horn [16].

In conclusion, although diagnosis might be difficult, multiple techniques may be reliable, as ultrasound, MRI, hysterosal pingogram and diagnostic hysteroscopy or laparoscopy, especially before a corrective surgical procedure is decided. An early diagnosis and surgical removal of these rudimentary cavitated masses will help relief symptomatic patients and avoid further complications.

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