MRI And USG Diagnosis Of Unicornuate Uterus With An Obstructed Rudimentary Horn: A Case Report

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Abstract

Mullerian anomalies have varying presentations, some of which overlap with a more common diagnosis. Pelvic pain represents one such presenting symptom and imposes considerable distress. Its broad range of etiologies, both gynecologic and non-gynecologic, makes pelvic pain a frustrating symptom for patients and physicians alike. Proper evaluation often may involve one or more diagnostic imaging modalities, and treatment may be medical or surgical. Though current imaging modalities such as MRI and 3D ultrasound are highly predictive of Mullerian anomalies, it is important to bear uncommon presentations of common pathologies in mind. Herein we report the case of a 16-year-old woman with severe menstrual pain whose 2D ultrasound and MR imaging suggested a unicornuate uterus with obstructed rudimentary horn.

Keywords: Mullerian anomaly, Unicornuate uterus, Magnetic resonance imaging (MRI)

Introduction

Congenital anomalies of the uterus and vagina (caused by alterations in development or fusion of the müllerian ducts) are associated with a high incidence of infertility and multiple obstetric problems. USG helps in the initial evaluation. MR evaluation allows accurate morphologic demonstration and classification of these anomalies and helps in planning an accurate treatment and also permits assessment of obstructive anomalies and associated pelvic and renal anomalies.

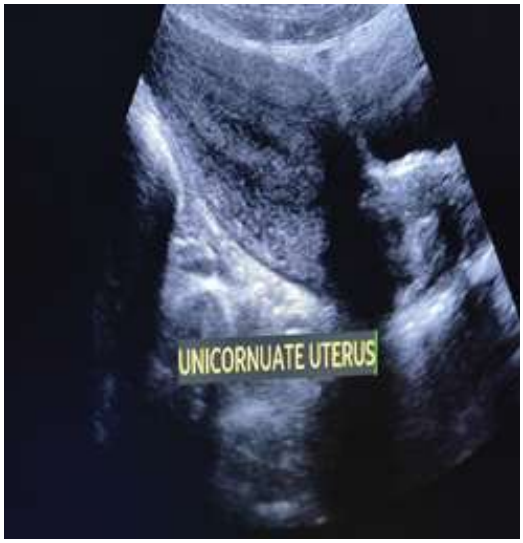
CASE:

A 16 year old unmarried girl who achieved her menarche at the age of 13 years with initial cycles of 28-30 days and menstrual bleeding lasting 2-3 days presented with history of severe menstrual abdominal pain. The initial sonography of the pelvis revealed a fluid-containing lesion abutting and deviating the uterine body and the fundus to the right. A detailed trans-abdominal USG was done and revealed a fluid-containing structure lying behind the urinary bladder and adjacent and on the left of a normal uterine body and fundus which appeared to be continuous with one cervix. Both the ovaries were identified separately and a provisional diagnosis of a congenital anomaly of the uterus (unicornuate) with obstruction and hematometra of the rudimentary horn was given. Transvaginal USG or HSG was not possible, as patient was unmarried. Patient was advised MRI pelvis to detect the cause of his problems. High-resolution MR imaging was performed using 1.5T Achieva system (Philips Medical Systems). MRI imaging protocol includes following: T2WI spin-echo sequence in

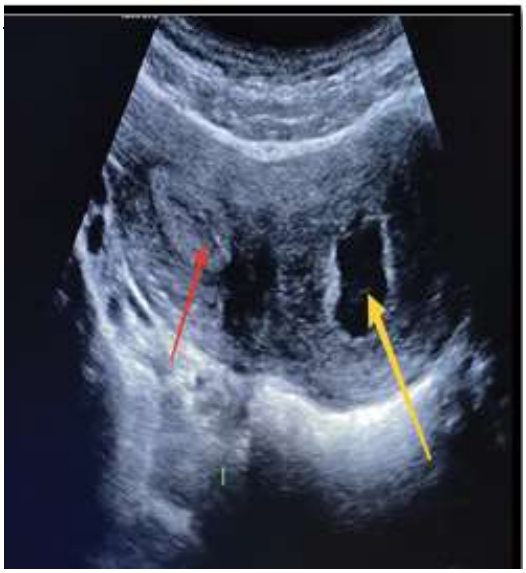
axial, coronal, and sagittal, FLAIR in the axial and coronal plane, VISTA sequence in axial and coronal plane, T1WI precontrast images in axial and coronal plane and T1WI post contrast images in axial and coronal plane. MR study showed continuity of uterus with on cervix. A globular structure measuring approximately 3.5 x 3.8 x 4.1 cm was seen lying to left side of uterus. Morphology of this structure was suggestive of uterine in nature with preserved zonal anatomy. Internal contents of the structure was hyperintense on T1WI ,T2WI and fat-suppressed images. Both the ovaries were normal and showed multiple small subcentimeter-sized follicles that were hyperintense on T2WI and hypointense on T1WI. Both kidneys and rest of major abdominal organs were normal. A diagnosis of congenital anomaly of the uterus – unicornuate uterus with left sided obstructed functioning rudimentary horn with a hematometra in the obstructed element was made.

Image 1:Ultrasound pelvis images of unicornuate uterus with an obstructed rudimentary horn. (a) Unicornuate uterus. (b) a unicornuate uterus (red arrow) with rudimentary horn on the left side containing a clot (yellow arrow). (c) rudimentary uterine horn with blood clot (yellow arrow).

(a)



(b)



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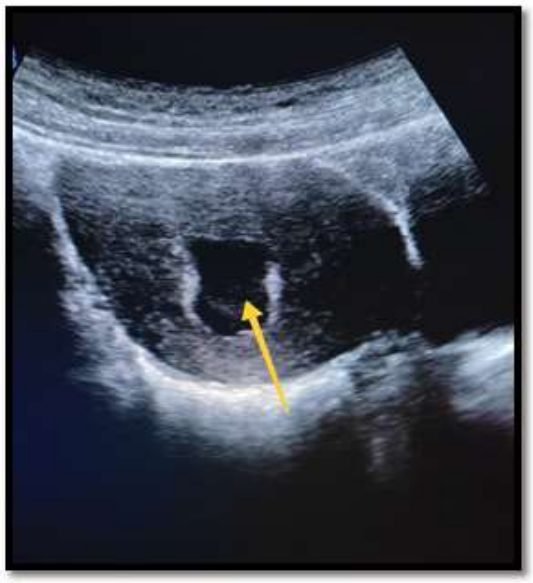
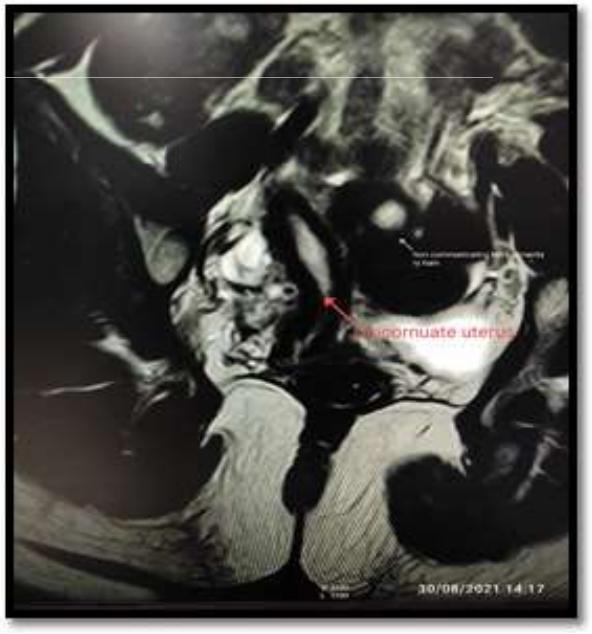


Image 2: MRI Pelvis (a). T2WI coronal section shows Unicornuate uterus with right uterine horn communicating with cervix (shown by red arrow) and left functional non communicating rudimentary horn containing collection hematometra likely(shown by white arrow). (b and c) T2WI axial sections shows a normal left and right ovary (Red arrow).

(a)



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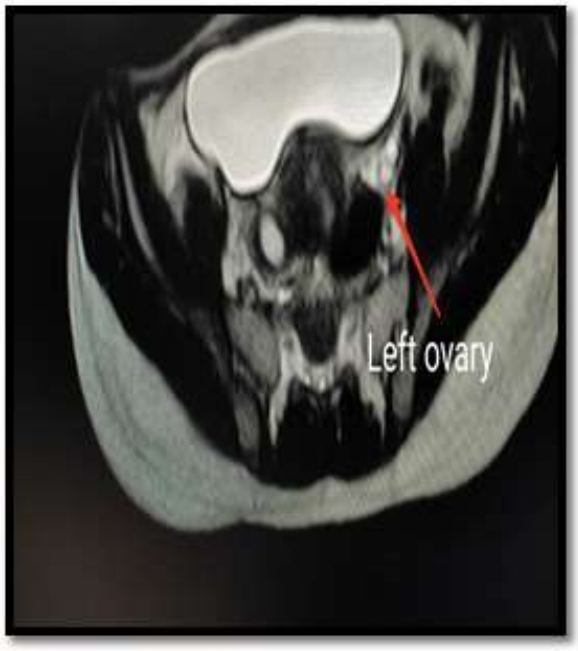


(c)

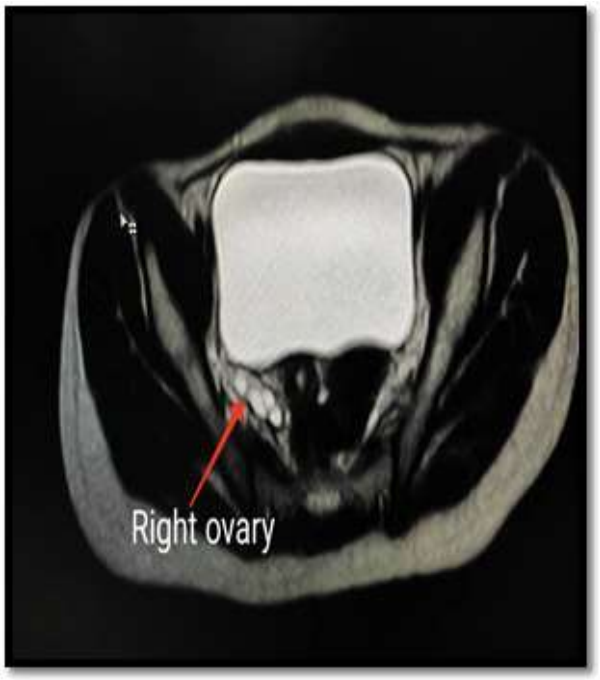
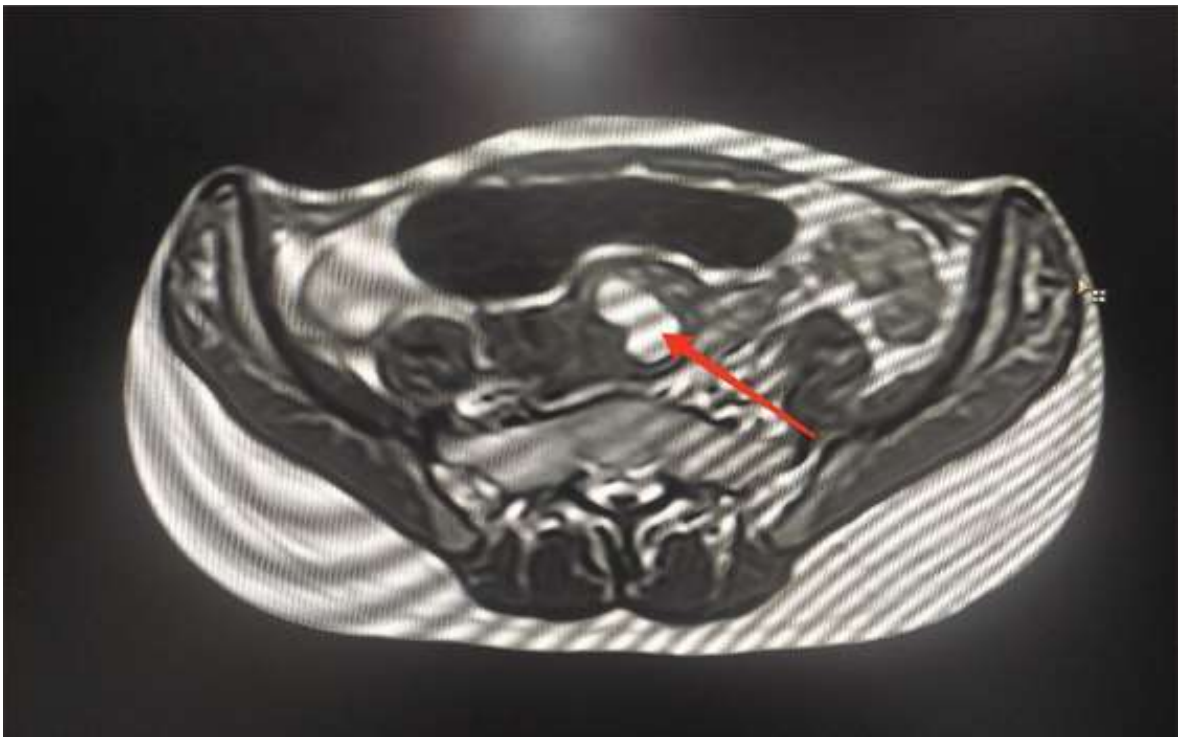
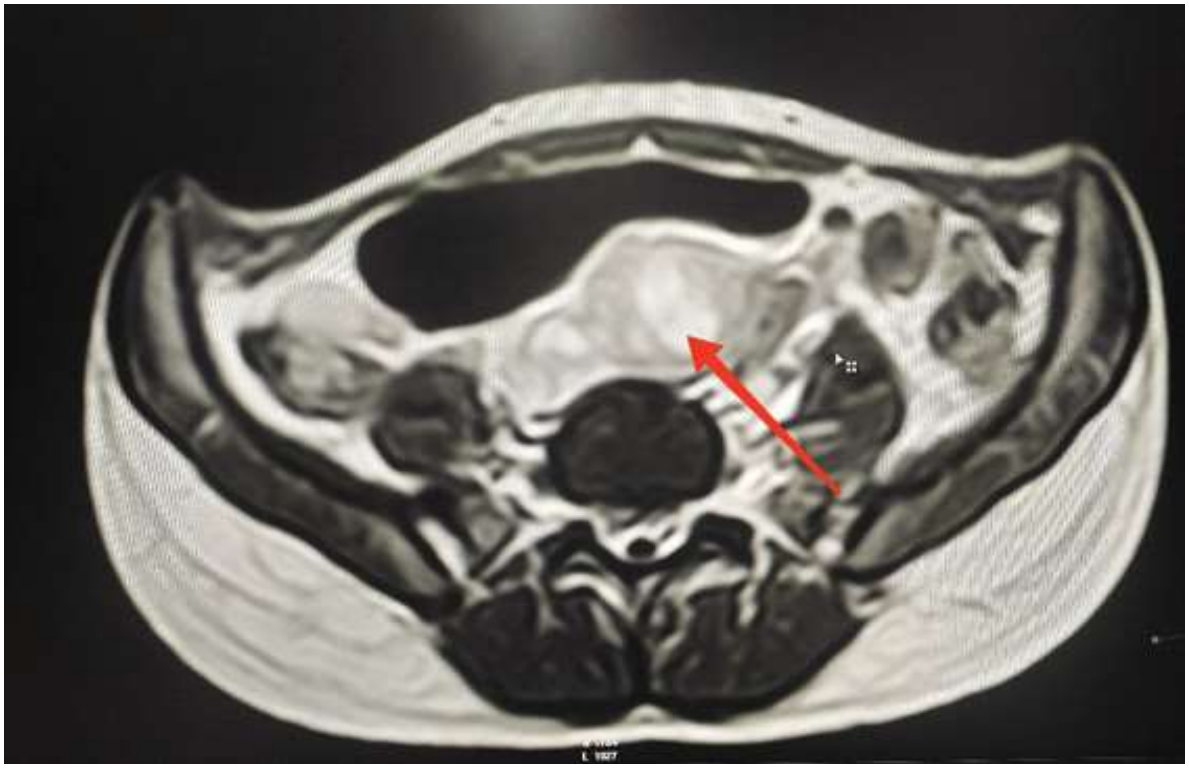


Image 3 :MRI Pelvis (a). T1W axial section shows unicornuate uterus with left rudimentary horn with hyperintense collection within s/o hematometra (red arrow). (b) VISTA axial section shows a unicornuate uterus with left rudimentary horn containing clot (red arrow).

(a)



(b)



Discussion

Fusion of mullerian ducts occurs normally between 6th and 11 weeks of gestation, with formation of uterus ,fallopian tubes ,cervix and proximal two third of vagina. Any disruption of mullerian duct development during embryogenesis results in congenital anomalies termed as mullerian duct anomalies. Distal third of vagina and ovaries originate from sinovaginal bud and primitive yolk sac respectively. Hence, mullerian duct anomalies are not associated with anomalies of external genitalia or ovaries.

Congenital anomalies of uterus or müllerian duct anomalies are not so common but they are treatable cause of infertility. They are estimated to occur in 0.1–0.5% of women, while the prevalence of these anomalies in females with a history of multiple pregnancy loss is about 3%. 25% of women with müllerian duct anomalies have reproductive problems like higher incidence of infertility, recurrent miscarriage, fetal malposition, preterm labor, endometriosis etc. Mullerian duct anomalies also associated with renal anomalies including renal agenesis, hypoplasia, ectopia, fusion, malrotation and duplication. Vertebral segmentation anomalies may also be associated with mullerian duct anomalies.[1-3]

Imaging plays an important role in diagnosis and treatment planning of mullerian duct anomalies. The first line investigation is sonography (abdominopelvic and trans-vaginal imaging if possible), because it is readily available , inexpensive and without ionising radiation ,However sonographic examination has its limitation as it is operator dependent and image resolution can be a limiting factor. Hysterosalpingography (HSG) allows visualization of the uterine cavity and fallopian tube patency. However, HSG does not provide information about external contour of uterus, hence characterization of müllerian anomalies can be difficult. MRI is considered the modality of choice for imaging uterine anomalies. MRI provides high-resolution images of the uterine body, fundus, cervix including external uterine contour and can assess the urinary tract for concomitant.[1-4]

Classification of anomalies

There is no universally acceptable mullerian duct anomaly classification system. Many classifications of uterine anomalies exist; like the American Fertility Society (AFS) classification, the modified AFS classification[5] by Rock and Adam[6] and the Buttram and Gibbons classification.[7]

According to the American Fertility Society (AFS) Müllerian duct anomalies are categorized most commonly into 7 classes, as below:

- Class I (hypoplasia/agenesis): This class includes agenesis or hypoplasia of uterus, cervix and proximal two third of vagina as part of- Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome and is extreme form of mullerian duct anomaly.
- Class II (unicornuate uterus): The unicornuate uterus results from complete/almost complete arrest of development of the müllerian duct. This anomaly has four subtypes: a) no rudimentary horn, b) rudimentary horn with no uterine cavity, c) rudimentary horn with communicating cavity to normal side, and d) rudimentary horn with a non communicating cavity.
- Class III (uterus didelphys): This anomaly results from complete failure of fusion of both müllerian ducts. The individual horns are fully developed, with duplication of uterine horns, cervix and proximal vagina.
- Class IV (bicornuate uterus): A bicornuate uterus results from partial nonfusion of the müllerian ducts. This anomaly is characterized by presence of a cleft (more than 1 cm in depth) in external uterine contour. The central myometrium may extend to the level of the internal os (bicornuate unicollis) or external os (bicornuate bicollis).

- Class V (septate uterus): A septate uterus results from failure of resorption of the septum between the two uterine horns (septum can be partial or complete). Differentiation between a septate and a bicornuate uterus is important because septate uterus is surgically correctable. Primary difference is appearance of external uterine contour- septate uterus has normal convex external uterine contour.
- Class VI (arcuate uterus): An arcuate uterus has a single uterine cavity with a convex or flat uterine fundus, the endometrial cavity, which demonstrates a small fundal cleft or impression.
- Class VII (diethylstilbestrol-related anomaly): The uterine anomaly was related in the female offspring of women exposed to DES during pregnancy. Female fetuses who are affected have a variety of abnormal findings that include uterine hypoplasia and a T-shaped uterine cavity. However, this anomaly is more of a historical interest and description of DES was stopped long way back.

The modified AFS classification by Rock and Adam: This classification correlates anatomic anomalies with embryologic arrests. Uterovaginal anomalies are categorized as dysgenesis disorders or vertical or lateral fusion defects. It is further subdivided into obstructive or nonobstructive forms. Immediate treatment is not needed for nonobstructive forms, but obstructive uterovaginal anomalies require immediate treatment because of retrograde flow of trapped fluids with increasing pressure on surrounding organs.

- Class 1—Dysgenesis of müllerian ducts. This class includes agenesis or hypoplasia of the müllerian duct derivatives (the uterus,cervix and upper two-thirds of the vagina).
- Class 2—Disorders of vertical fusion. There is failure of fusion of the müllerian system with the sinovaginal bulb (cervical dysgenesis and obstructive and nonobstructive transverse vaginal septa).
- Class 3—Disorders of lateral fusion. It comprises of a duplicated or partially duplicated reproductive tract. The disorders are due to impaired fusion and/or septal resorption of fusing müllerian ducts attempting to form the uterus, cervix, and upper vagina. It includes anomalies due to failure of fusion of the paired müllerian ducts (as in didelphic and bicornuate uteri) and failure of midline septum resorption after fusion (as in septate uterus). Disorders due to lateral fusion defects are further subclassified into (a) the symmetric nonobstructive form (unicornuate, bicornuate, didelphic, septate, and DES-related uteri) (b) the asymmetric obstructive form (unicornuate uterus with obstructed horn, double uterus with unilaterally obstructed horn, and double uterus with unilaterally obstructed vagina).
- Class 4—Unusual configurations and combinations of defects.

The case we described was unicornuate uterus with an obstructed rudimentary contralateral horn and it was classified as Class II(d) of the American Fertility Society (AFS) Classification Scheme or Class 3 of the modified AFS classification by Rock and Adam.

Conclusion

In conclusion, diagnosis of mullerian duct anomaly is important because of high associated risk of infertility, abortion and endometriosis. Ultrasound is initial modality of investigation. MRI is modality of choice, as it shows accurate morphology, allows classification of mullerian duct anomalies and helps in treatment planning.

Declaration of Patient Consent: Written informed consent was obtained from patient for publication of this report.

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Conflict of interest: Nil

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