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Case Report

Magnetic Resonance Imaging and Ultrasound Depiction of Robert's Uterus: A Rare Müllerian Duct Anomaly Causing Left Hematometra and Left Hematosalpinx

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Abstract

Robert's uterus is a rare unique mullerian duct anomaly, first reported by Robert in 1970. The uterus shows two uterine cavities with one is being blind causing hematometra and the other cavity is communicating to a single cervix. So the patients present with cyclic abdominal pain during menstruation in spite of normal menstrual flow.

Any associated hematometra or endometriomas complicating this condition can be depicted with MRI which aid in the appropriate management.

We present magnetic resonance imaging (MRI) and ultrasound findings in a case of Robert's uterus in 16 years old young woman.

Keywords: *Dysmenorrhea; Hematometra; Mullerian Duct Anomaly; Robert's Uterus; Septate Uterus*

Background

There are various forms of mullerian ducts anomalies and each anomaly is distinctive. In general population, the incidence of müllerian duct anomalies is 0.1 - 3.5%, but they are much higher in patients with recurrent pregnancy loss: 5 - 10%. The incidence has been increased due to the availability of better imaging facilities [1].

American Fertility Society (AFS) classification organizes müllerian duct anomalies according to major uterine anatomic defects and allows for standardized reporting methods. The most common anomaly is septate uterus (AFS Class V). It is resulting from incomplete resorption of the medial septum after complete fusion of the müllerian ducts has occurred. There are various forms of septate uterus like complete, partial and segmental. The complete form extends from the fundal area to internal os and divides the endometrial cavity into 2 components and may be associated with a longitudinal vaginal septum. The patient commonly presented with dysmenorrhea, dyspareunia, primary or secondary infertility, pregnancy loss and obstetric complications [2].

Robert's uterus is an extremely rare müllerian duct anomaly, characterized by asymmetrical septum dividing the uterus into two cavities, one cavity is obstructed. The other one communicates normally with the single cervix, which is responsible for cyclical menstrual flow. The obstructed cavity has functional endometrium, so its secretions become retained with every menstruation and therefore the patient presents with cyclical pain [3].

Herein is described the case of a 16 year-old virgin single woman whose Robert's uterus was diagnosed by MRI and ultrasound studies.

Methods

We report ultrasound and MRI findings in a young women with Robert's uterus who presented with cyclical menstrual pain and dysmenorrhea.

Case Report

A 16 year old single virgin female presented with dysmenorrhea with pain and fullness on the left side of the pelvis, and she has normal menstrual flow. She is a known case of caudal regression syndrome with imperforate anus. She underwent multiple abdominal surgeries between 2001 and 2006 for colostomies, repair of fistulas, other genitourinary pathologies as well as peritoneal wash. She has stopped using colostomy and now passing stool normally. She is using self-catheterization for urinary incontinence.

On the abdominal examination, fullness and tenderness was seen in the left flank.

Transabdominal Pelvic Ultrasound scan (Figure 1) suggested bicornuate uterus with heterogeneous collection in the left horn (Figure 1a). Normal empty right sided uterine cavity (Figure 1b). A left-sided, extra uterine complex cystic lesion suggestive of left hematosalpinx (Figure 1c). Normal left ovary is seen (Figure 1d), excluded the possibility of left endometriotic cyst. Both kidneys were normally visualized with minimal dilatation of their pelvicalyceal systems.

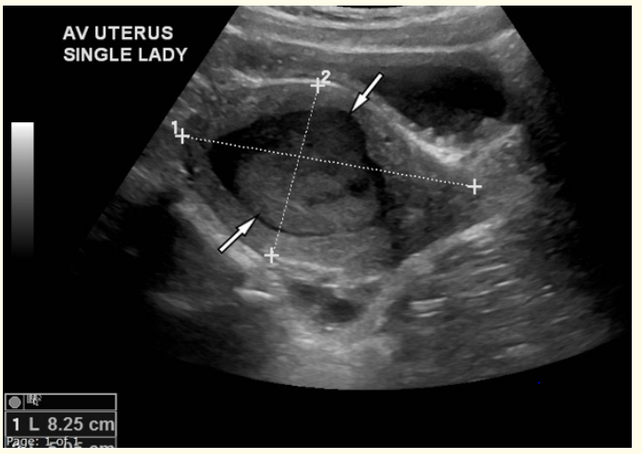
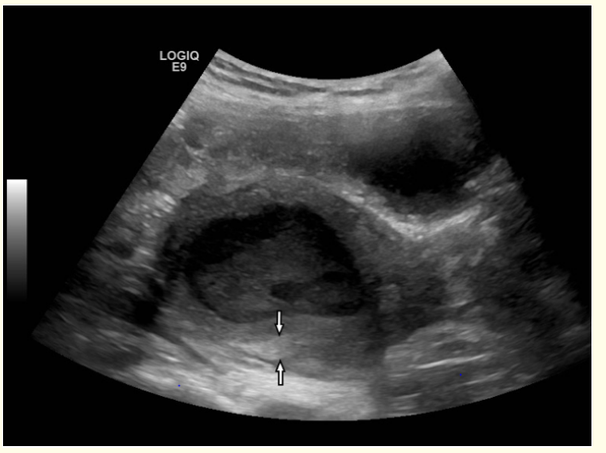


Figure a



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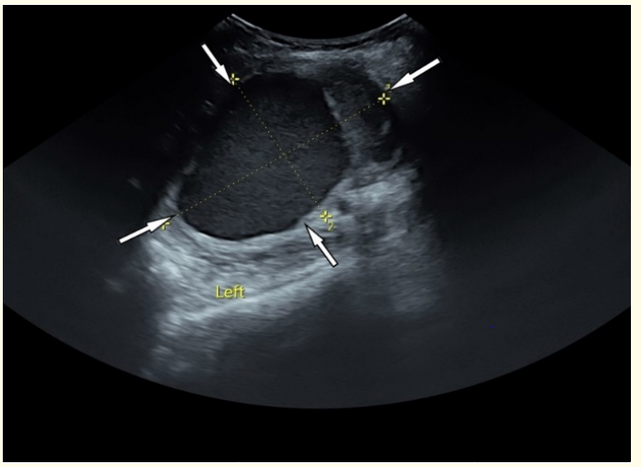

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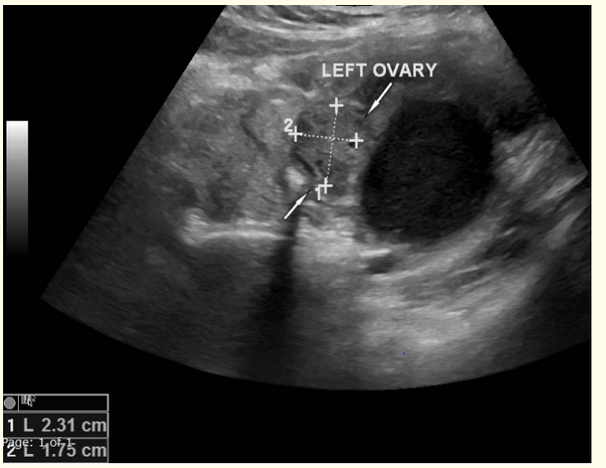
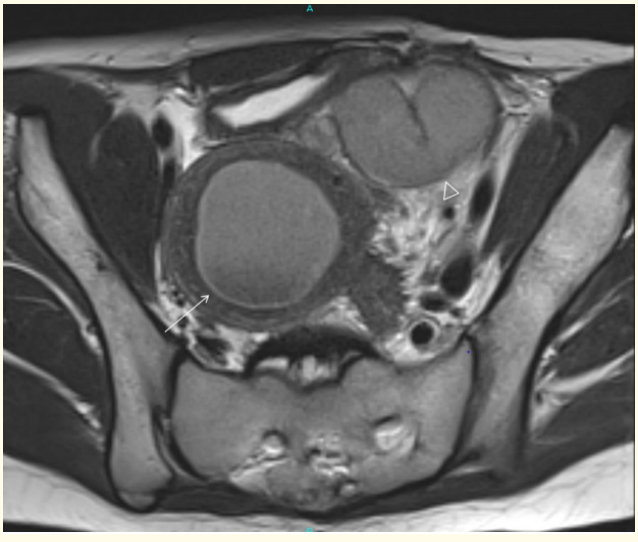


Figure d

Figure 1a: *Distended left uterine cavity (measures: 8.3 x 5.1 cm) with heterogeneous collection of blood products (arrows).*
Figure 1b: *Normal empty right sided uterine cavity (arrows).*
Figure 1c: *left hematosalpinx (arrows). Markedly distended left fallopian tube (measures: 9.5 x 6.3 cm) with partial septum and heterogeneous collection of blood products.*
Figure 1d: *Normal let ovary (arrows). It measures: 23 x 18 mm and is seen mildly displaced medially by large left hematosalpinx.*

Figure 1: *Trans abdominal Pelvic Ultrasound.*

MRI was performed in Avanto 1.5T system (Siemens Medical Systems) showed asymmetric septate uterus with normal smooth its outer fundal contour. The left-sided cavity was obstructed with associated hematometra. Transverse (Figure 2a-c) and coronal (Figure 3a-c) T2-weighted MR images of the pelvis show obstructed left horn of the uterus filled with hemorrhagic fluid as well as left hematosalpinx and both of them are not communicating with the normal right uterine horn or the cervix. Transverse T1W fat-saturated image (Figure 2d), shows hyperintense signal of the obstructed uterine horn and left hematosalpinx.



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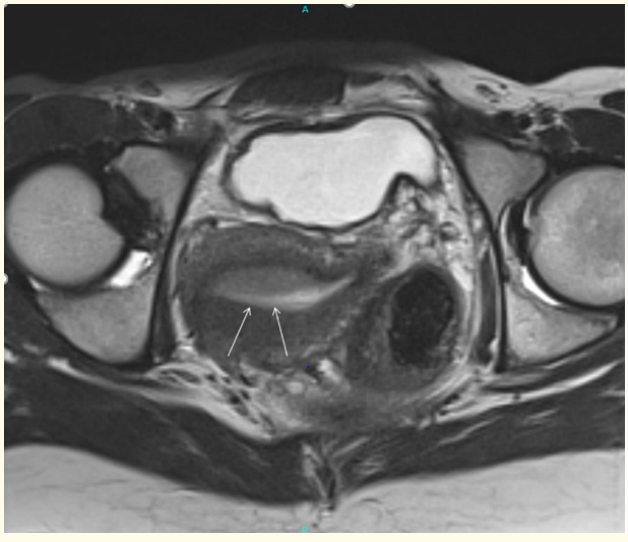

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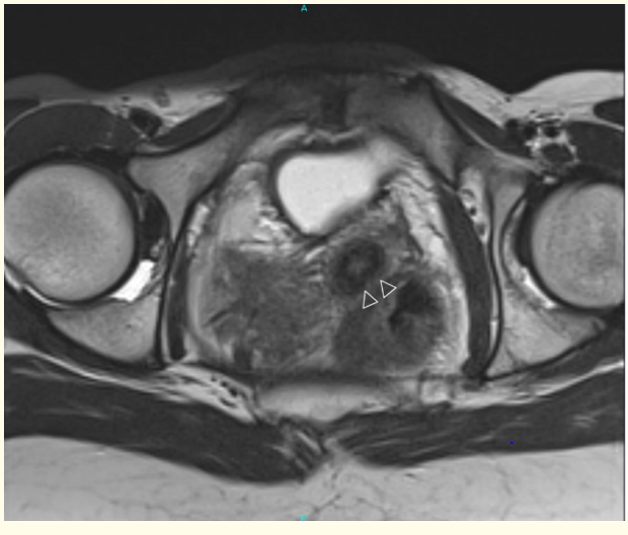

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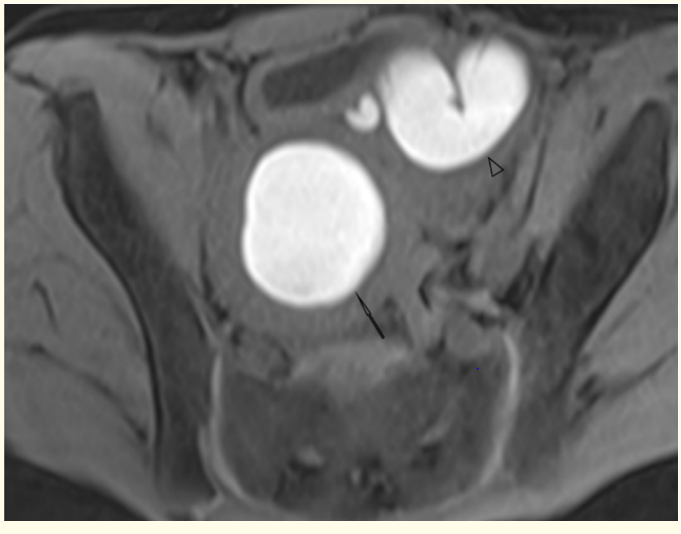

Figure d

Figure 2a to 2c: *Transverse T2-weighted MR images of the pelvis show an obstructed left horn of the uterus filled with hemorrhagic fluid which shows low signal intensity (white arrow) associated with left hematosalpinx (white arrowheads), both of which are not communicating with the normal right uterine horn (double white arrows) or the cervix (double white arrowheads).*

Figure 2d: *Transverse T1W fat-saturated image, shows hyperintense signal of the obstructed uterine horn (black arrow) and left hematosalpinx (black arrowhead).*

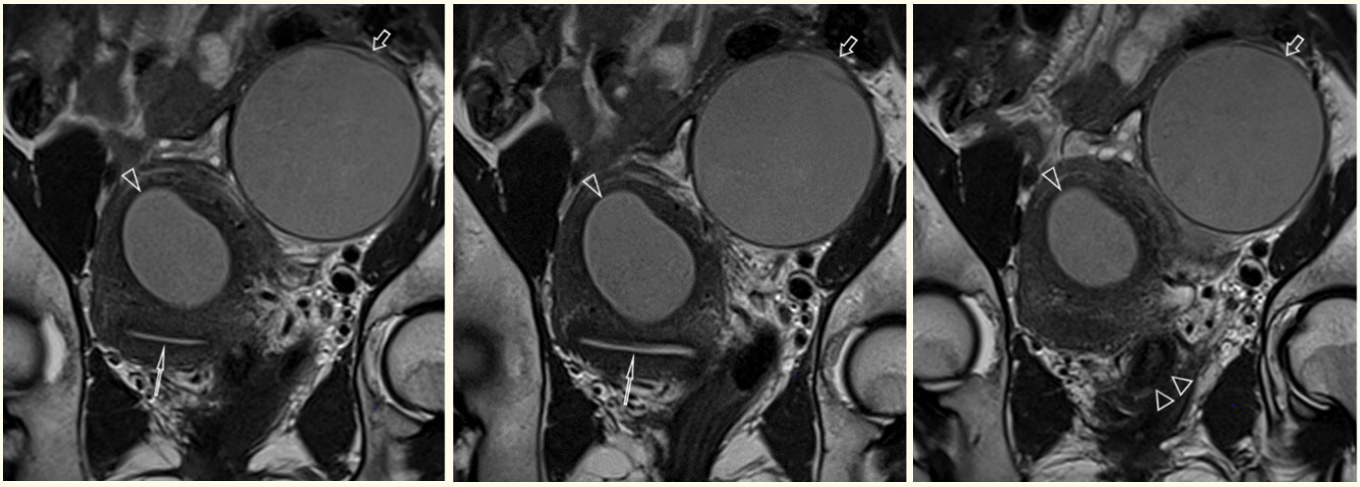

Figure a *Figure b* *Figure c*

Figure 3a to 3c: *Coronal T2-weighted MR images of the pelvis show an obstructed left horn of the uterus filled with hemorrhagic fluid which shows low signal intensity (arrowhead) associated with left hematosalpinx (short arrow), both of which are not communicating with the normal right uterine horn (long arrow) or the cervix (double arrowheads). Normal smooth outer fundal contour of the uterus is also noted.*

MRI study also revealed polycystic configuration of the ovaries, caudal regression syndrome with partial sacral agenesis and partial block vertebra at L2-L3, neurogenic bladder with diversion stoma at the anterior abdominal wall and left hydroureter.

Patient was consulted about need of hysteroscopy to relive her pain and symptoms. Patient refused to spare her virginity, other options will be discussed with her later on.

Discussion

Robert's uterus is a unique congenital müllerian anomaly, it is an asymmetric septate uterus, first reported by Robert in 1970 [4]. Very few cases have been reported in literature, so far [5,6].

It is a variant of septate uterus with oblique course of the inferior part of the septum resulting in obstructed hemi-uterus with one blind horn causing hematometra and the other hemi-uterus is communicating with a single cervix and preserved normal extrauterine morphology. The functioning endometrium in the obstructed cavity releases cyclical secretions which can extend into the fallopian tube and the adnexal region resulting in formation of hematosalpinx and endometriomas. Therefore, the patients classically present with cyclical abdominal pain and dysmenorrhea [3].

The other müllerian duct anomaly; noncommunicating rudimentary horns usually present after menarche with development of hematometra, hematosalpinx or endometriosis resulting in progressive abdominal pain and dysmenorrhea. The mean age of gynecologic horn presentations is 23 + 7.4 years and median duration of symptoms is 24 months before presentation [4]. Our patient presented at 16 years of age.

Associated hematometra, hematosalpinx and adnexal endometriomas can be visualized by Ultrasound. However, sonography is limited in differentiating between different subtypes of müllerian duct anomalies.

The modality of choice for full visualization of the extent of the anomaly and any associated complications is MRI. It provides perfect anatomical detail and direct visualization of the uterine contour, especially on coronal images [3].

MRI provides excellent tissue characterization for differentiating septate from bicornuate uterus. MRI Coronal T2W images are ideal for demonstrating asymmetric uterine septum dividing the endometrial cavity along with the blind ending cavity and distended with hematometra. Chronic blood products typically appear bright signal on T1W and T1W fat-suppressed images with a shading effect on T2W images. T1W fat-sat images are very sensitive in detecting small endometriomas [7].

The treatment of Robert's uterus is not fully established, because it is reported very rarely in the literature, as case reports. Open and minimal invasive surgical procedures have been described. The aim of treatment is drainage of the hematometra, prevention of its recurrence and drainage/excision of adnexal endometriomas. The recurrence of hematometra can be avoided by complete excision of the obstructed cavity with preservation of the normal cavity or by ablation of the functional endometrium in the obstructed uterine cavity or by unification of both uterine cavities by incising the septum. Open laparotomy may be a better approach if there is associated adnexal endometriomas and adhesions, as adequate clearance of adnexal endometriomas can be done using open surgery. In both hysteroscopic and laparoscopic approaches, incision of the septum has been described with good outcomes. This can keep more possibilities for improving the uterine cavity for a better reproductive outcome later on and postpone the irreversible procedures [3].

Conclusion

Robert's uterus is a very rare müllerian duct anomaly, which can be better evaluated with MRI than ultrasound. This case emphasizes that müllerian duct anomalies should be considered in the differential diagnosis of cases with severe dysmenorrhea in spite of normal menstruation. The diagnosis remains a challenge to the clinicians. With early diagnosis, minimal invasive procedures may be more effective, before the formation of hematosalpinx or adnexal endometriomas.

Conflict of Interest

The authors declared no conflict of interest.

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