The Wandering Twin: A Case of a Uterine Didelphys with the Obstructed Hemiuteri in the Anterior Abdominal Wall

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Mullerian duct anomalies (MDAs) are congenital defects of the female genital system that arise from abnormal embryological development of the Mullerian ducts. A didelphys uterus, also known as a "double uterus," is one of the least common amongst the MDAs. Reported here is a case of a 16 year old female with a uterus didelphys with the obstructed left hemiuteri adherent in the anterior abdominal wall, and an endometriotic cyst on the same side. She underwent hysteroscopy-guided vaginoscopy, laparoscopic left hemihysterectomy, left oophorocystectomy. Cases such as these require careful preoperative planning and diagnostic imaging for more accurate diagnosis and, hence, for the most appropriate surgical procedure to be carried out. 3D ultrasonography and Magnetic Resonance Imaging have been the most widely used imaging techniques. The goals of management are to relieve the symptoms of obstruction and to restore the normal anatomy as much as possible in order to provide the best chance for future fertility.

Key words: mullërian duct anomalies, uterine didelphys

Introduction

Developmental anomalies of the mullerian ducts are some of the more fascinating and challenging in the field of Obstetrics and Gynecology. Uterine malformations make up a heterogeneous group of congenital anomalies that can result from the underdevelopment of the Müllerian ducts, disorders in their fusion and/or alterations in septum resorption. The prevalence of uterine malformations is difficult to establish. Incidence rates vary widely and depend on the study. Most authors report incidences of 0.1-3.5%.[1,2,3,4] In 2001, Grimbizis and colleagues reported that the mean incidence of uterine malformations was 4.3% for the general population and/or for fertile women. The discrepancy among different publications stems from their use of different diagnostic techniques, heterogeneous population samples and the clinical diversity of Mullerian anomalies.[5]

Most müllerian duct anomalies (MDAs) are associated with functioning ovaries and age-appropriate external genitalia. These abnormalities are often recognized after the onset of puberty. In the prepubertal period, normal external genitalia and age-appropriate developmental milestones often mask abnormalities of the internal reproductive organs. After the onset of puberty, young women often present to the gynecologist with menstrual disorders. Late presentations include infertility and obstetric complications.

Due to the complexity of presentations, diagnosis of Müllerian malformations requires the use of more than one imaging method in 62% of the cases.[7,8] Hysterosalpingography (HSG) is the method traditionally used to evaluate the cervical canal, uterine cavity and fallopian tubes. Its

efficacy in diagnosing anomalies is debatable and varies according to the specific type of malformation. Ultrasonography has a sensitivity of around 44%, varying according to the specific type of malformation under evaluation, the patient's body composition, the radiologist's experience and the type of transducer used. Transvaginal ultrasonography allows a more detailed analysis of the endometrium, uterine cavity and cervix. The specificity of this examination ranges from 85 to 92%.[9,10,11] Recently, three-dimensional ultrasonography has shown high specificity and sensitivity in evaluations on all uterine anomalies, including Müllerian malformation. The specificity of magnetic resonance imaging (MRI) ranges from 96 to 100% for diagnosing Müllerian malformations.

Because of the wide variation in clinical presentations, müllerian duct anomalies may be difficult to diagnose. After an accurate diagnosis is rendered, many treatment options exist, and they are usually tailored to the specific müllerian anomaly. Restoration of normal uterine architecture and preservation of fertility are the goals of surgical treatment of uterine anomalies.

The Case

A 16 year old female sought consult at our institution due to persistent left lower quadrant pain. She had her menarche at 12 years of age, but regular monthly intervals only started one year ago, with accompanying progressive dysmenorrhea relieved by intake of pain relievers. Past history includes an inguinal repair and excision of ectopic left kidney done in 2011.

Two weeks prior to consult, she experienced severe left lower quadrant pain and was rushed to the emergency room, where a palpable mass was felt on abdominal examination. She was subsequently given unrecalled IV pain medications, which afforded temporary relief. She was discharged with orders to seek gynecologic consult.

Upon her visit to the gynecologist, a transrectal ultrasound was done and showed a probable uterine didelphys with hematometra on the left; Endometriotic cyst, Left (Figure 1).

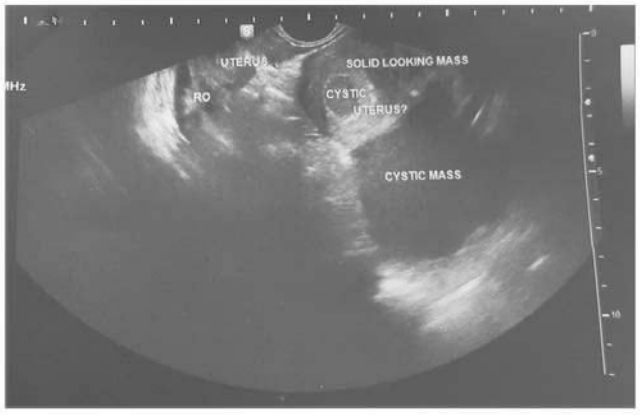

Figure 1. Transrectal ultrasound showing a normal uterus on the right, normal right ovary, with a solid looking mass measuring 4.7cm x 4.6cm x 4.2cm containing a cystic inner component (hemiuteri with hematometra?), and an endometrial cyst on the left measuring 8cm x7cm x 7cm.

She was referred to our institution. Abdominal physical findings reveal an 8cm x 8cm x 6cm tender, firm to doughy mass at the left lower quadrant, with limited mobility. External genitalia was normal with Tanner 3 staging of pubic hair, hymen was annular and intact. On rectal exam, the cervix was firm and deviated to the right with a normal-sized uterus; to the left of the cervix was the inferior pole of a firm to doughy mass measuring approximately 5cm in diameter.

Working impression was a Uterine Didelphys with Obstructed Hemivagina (OHVIRA syndrome-in the background of an ipsilateral ectopic kidney) versus a Unicornuate Uterus with a non-communicating rudimentary horn.

MRI showed bicornuate uterus, possibly bicornis bicollis type with hematometra, left; Hydrosalpinx, left and large ovarian cyst, left. (Figures 2 & 3)

She was then admitted and underwent hysteroscopy-guided vaginoscopy, with laparoscopic left oophorocystectomy and left hemihysterectomy. Endoscopic vaginal exploration showed smooth and pink vaginal walls with presence of only one cervix. Laparoscopy revealed a normal-sized right hemiuterus with pink and smooth serosal surface. The ipsilateral fallopian tube and ovary were grossly normal. A band of fibrous tissue directly inferior to the bladder was identified connecting the normal right

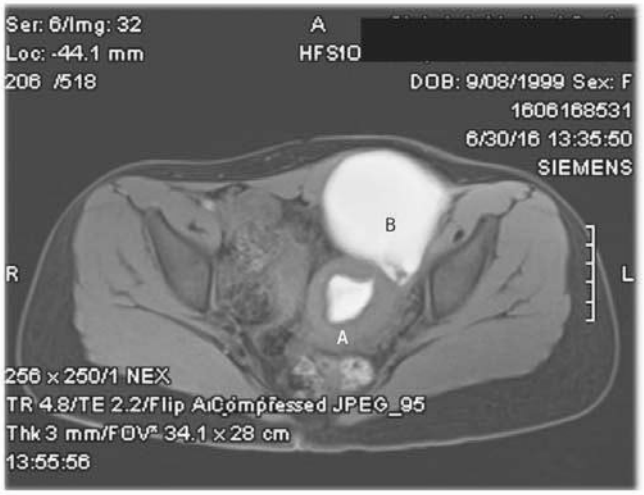

Figure 2. MRI with contrast showing a probable left uterine horn with hematometra (A) and ovarian cyst (probably endometrial cyst) on the left.

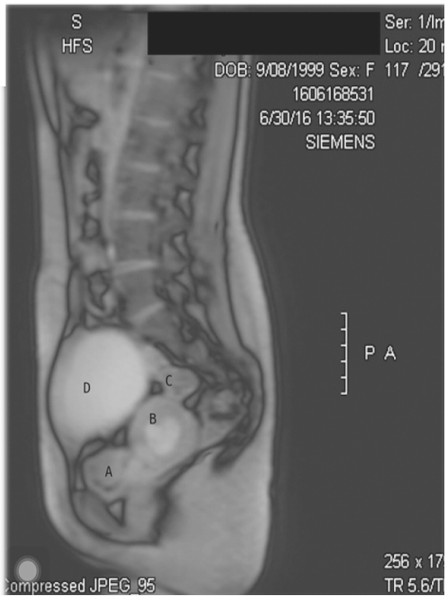

Figure 3. Showing lack of fusion of the uterine corpus extending inferiorly into a single vaginal canal. The external uterine contour is concave with widely divergent uterine horns. The right uterine horn (A) is smaller than the left uterine horn (B). There was also note of hydrosalpinx (C) and an ovarian cyst (D) on the left.

hemiuterus to the left hemiuterus, which was completely enclosed within the parietal peritoneum. An 8cm x 6cm x 6cm cystic, thin-walled mass was noted attached to the left hemiuterus. It contained 100cc of thick chocolate-brown fluid. The left fallopian tube could not be identified due to dense adhesions. The appendix and liver surface was grossly unremarkable. Upon morcellation, the left hemiuterus was seen dilated by blood from a functional endometrial lining. The cervical canal was smooth. Postoperative course was unremarkable, and she was discharged on the second postoperative day. Histopathology showed leiomyoma, secretory endometrium, endometriotic cyst, fallopian tube endometriosis and hematosalpinx.

She was seen at the outpatient clinic on her 1st day of menstruation with no dysmenorrhea. She was put on cyclic combined oral contraceptive pills. Final diagnosis was Uterine didelphys with obstructed functional left hemi-uterus, and endometriotic cyst, left ovary.

Discussion

Uterine didelphys is an embryonic malformation of the female genitourinary system that occurs between the 12th and 16th week of gestation. It is caused by non-absorption of the septum formed as a result of these 2 mullerian ducts. It is composed of two separate normal-sized uteri; more commonly, the two separate cervix uteri are fused at the lower uterine segment.[12] In cases of outflow obstruction, it commonly occurs because of a concomitant blind hemivagina, hence the patient presents with a bulging mass at the vagina and on ultrasound or MRI would show hematometra with hematocolpos. However, in this patient, the obstructed hemiuterus was located outside of the pelvic cavity, so she did not present with hematocolpos on both imaging studies done. Intraoperatively, the left hemiuteri was seen at the left lower quadrant enveloped by parietal peritoneum. The cervix of this hemiuteri was blocked by the anterior abdominal wall, which caused the outflow obstruction. Based on literature review, there were no similar cases on this unusual location of the hemiuteri. There was one case reported by Yoshiki, et al. in 2007 of a uterine didelphys presenting in the retroperitoneum. This patient presented with an obstructed hemivagina

and ipsilateral renal agenesis; laparoscopic resection of the right hemiuterus was performed.[12]

The manifesting symptoms usually appear only after menarche and consist of dysmenorrhea, severe abdominal pain, and the presence of a pelvic or intraabdominal mass.[13] For this case, the symptoms appeared about 3 years after menarche, when her menses started to occur regularly in interval and flow. The incidence of endometriosis in mullerian anomalies occurred in 19.8% of patients in one study group compared to controls (patients with no mullerian duct anomalies).[14] However, if we take into consideration only patients with mullerian duct anomalies, endometriosis was present in ten of 13 women with functioning endometrium, patent tubes, and outflow obstruction, whereas it could be identified in only 16 of 43 women with no obstruction (77 versus 37% respectively).[15]

The embryological development of the female reproductive system is closely related to the development of the urinary system, and anomalies in both systems may occur in up to 25% of these patients, namely: renal agenesis and dysplasia, double collecting system, and ectopic kidney or ureter.[15] They are seen on the same side of the mullerian defect. This was exemplified in this case, for the patient had an ectopic kidney on the left as an incidental intraoperative finding during herniorrhaphy in 2011. Other associated malformations may affect the gastrointestinal tract (12%) or musculoskeletal system (10-12%).[16]

This case is of particular significance given that uterine didelphys is one of the least common malformation of the mullerian ducts, but carries the best prognosis in terms of alleviation of symptoms and future fertility. Unicornuate and didelphys uterus have term delivery rates of ~45.[17] The fertility of women with untreated didelphys uterus has been shown by some sources to be better than those with other Mullerian duct abnormalities but still less than women with normal uterine anatomy. There is also an increased risk of spontaneous abortion, fetal growth retardation, and prematurity with an estimated 45% (or lower) chance of carrying a pregnancy to term in comparison to a normal uterus, which is similar to that of a unicornuate uterus. This indicates poor reproductive performance, but still not as poor as a septate or bicornuate uterus which are more common amongst the Mullerian duct anomalies.18 Hence, it is of utmost importance that an accurate diagnosis be made at the soonest possible time. There are several techniques available for the evaluation of uterine malformations. When the cavity only is to be assessed, hysterosalpingography (HSG) and hysteroscopy are especially useful. Laparotomy and laparoscopy can be also used for examination of the uterine fundus. There are two techniques, however, that combine the study of both these structures, which is indeed relevant for the diagnosis: magnetic resonance imaging (MRI) and three-dimensional (3D) ultrasound. While MRI is a useful option in the diagnosis of Mullerian anomalies, with numerous studies having proved its excellent efficacy in this field. 3D ultrasound represents a valid alternative, because, in addition to its lower cost and better tolerance by patients, it provides images of very similar quality to those yielded by MRI. In this study, the authors have concluded that there is a high degree of concordance between 3D ultrasound and MRI in the diagnosis of uterine malformations, the relationship between cavity and fundus being visualized equally well with both techniques. The adnexae can also be well visualized in both imaging techniques.[19]

The treatment for Müllerian anomalies varies according to the specific type of malformation found in each patient, and whether the patient is symptomatic. With careful pre-operative considerations and surgical technique, laparoscopic hemihysterectomy for obstructed uterine didelphys is safe, feasible, and gains all the benefits of a minimally-invasive approach.[20]

Patients with an obstructed uterine horn are at increased risk of endometriosis, but the endometriosis usually resolves after the removal of the obstructed hemiuterus. Excision of the obstructed rudimentary blind horn will prevent endometriosis by eliminating reflux, and will also prevent development of a pregnancy (and pregnancy complications) in the obstructed uterine horn.[21] The authors of this paper recommended

that the obstructed rudimentary noncommunicating uterine horn should be removed laparoscopically.[22]

Management of patients with müllerian agenesis includes psychosocial counseling to address the functional and emotional effects of genital anomalies as well as correction of the anatomical defect. After the diagnosis of müllerian agenesis, the adolescent should be offered counseling to emphasize that healthy sexual relationships are possible. Future fertility options should be addressed with adolescents and their parents or guardians.[23]

Conclusion

Uterine didelphys presenting with outflow obstruction is a well established entity, as it has been described as early as 1922. Although relatively uncommon, it is frequently described in literature because of its dramatic presentation.12 The importance of imaging for these kinds of anomalies cannot be overemphasized; the surgical procedure will depend on a careful preoperative and accurate diagnosis. Surgery is done for patients with symptoms, more commonly due to outflow obstruction, and the type of surgery will depend on the type of anomaly present. Studies have recommended laparoscopy to be the treatment of choice, considering that most of these patients are adolescents or in the early reproductive years, and would most benefit from a minimally-invasive procedure in terms of future fertility chances and long-term reduced chance of postoperative adhesions. Aside from the surgical treatment, patients should also be thoroughly counseled in the emotional and psychological aspects of their condition.

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