

Unusual management of a non-communicating uterine horn

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We report a case of a 15-year-old young single Saudi female, who presented with very scanty periods, increasingly of severe disabling dysmenorrhea and abdominal swelling. Her condition worsened over the last few months before presentation. The patient was clinically examined and fully investigated. An 18×8cm non-communicating right uterine horn with a small uterine horn connected to the outflow tract was found on investigation. In view of her age and general condition she was managed using a conservative surgical approach, rather than the standard radical excision of the uterine horn. The aim was to ensure a regular proper period in the future, which was achieved at 18 months of follow-up.

Case

A 15-year-old single Saudi female presented to our hospital complaining of abdominal distension, with increasing severe dysmenorrhea for the previous 2 years. In the past her menarche was omitted at age 12 years and she had a regular menstrual cycle every 28 days, with menstrual flow lasting for 4 days. Bleeding was extremely scanty with disabling dysmenorrhea that did not respond to injectable non-steroidal, anti-inflammatory analgesics. Her dysmenorrhea got worse over the previous one year with lower abdominal swelling. No gastrointestinal tract or urinary symptoms were observed. The patient's medical, surgical, developmental and family histories were unremarkable.

On clinical examination, her weight was 55 kilograms, height 160 centimeters, and blood pressure was 120/70 mm Hg. Her general physical examination was normal. Secondary sexual characteristics were normal. The lower abdominal examination revealed a mass, which was felt extending from the pelvis to the right lower quadrant and hypogastrium. The mass was mobile, very tender, soft and measured approximately 16 x18 cm in widest diameters.

Rectal examination confirmed the presence of a 20-cm pelvi-abdominal mass, which was firm, slightly mobile and tender. The complete blood count and renal function tests were normal. An abdominal and pelvic ultrasound reported normal abdominal organs but an absent right kidney, with a pelvic mass measuring 18×16 cm that might have represented a pelvic kidney. Intravenous pyelogram using an I.V. injection of 50 mL of Ominopaque 300 mg/mL revealed a normal left kidney with no opacification of the right kidney until the end of the study. No enhancement of the pelvic mass was detected. It was suggested that there was agenesis of the right kidney. A contrast-enhanced CT of the abdomen and pelvis confirmed the absence of the right kidney and the presence of a cystic structure in the lower abdomen most likely representing an isolated non-communicating uterine horn measuring 18×18 cm. A smaller horn measuring 3×2 cm was seen communicating with the vagina. A small collection of an unidentified source was also

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seen in between the two horns. Figures 1 and 2 are images selected from an enhanced CT scan of the pelvis showing a large cystic structure (18×18 cm) with relatively dense fluid and smooth wall, which displaced the urinary bladder anteriorly. The upper CT sections show the previously described cystic mass with two soft tissue structures (horns) attached to it, the larger being in the right side with a visualized cavity filled with fluid. These represent a small uterine horn and a huge hematocolpus.

An isotope study performed after IV injection of technetium-DMSA revealed the presence of a single left kidney. There was no evidence of renal tissue in the pelvis and the mass seen on previous CT was most likely hematocolpus (Figure 3).

The patient and her parents were counseled and consent was obtained for surgery. She was taken to the operating room, and cystoscopy was performed in a lithotomy position using a 20 French cystoscope. This showed normal healthy mucosa of the urinary bladder with severe mass pressure effect posteriorly, a patent left urethral orifice with normal urine flow and a completely absent right urethral orifice. The patient was repositioned to the supine position after inserting a Foley's catheter in the urinary bladder, and the abdomen was opened through a low transverse incision (Pfannenstiel incision). Laparotomy findings were as follows: A huge 18×18 cm right non-communicating uterine horn with a hugely dilated right fallopian tube (right hydrosalpinx) measuring 10×8 cm. The right and left ovaries were visualized and appeared grossly normal. A normal left fallopian tube was connected to the left horn. In view of the very small left uterine horn and her history of scanty periods, a decision was made against a right hemihysterectomy, which is the usual management in such cases, and it was decided to perform conservative surgery. The mass was incised at its medial aspect in a circular manner with a diameter of approximately 3 cm. Six hundred mL of altered dark blood were drained, the cavity was curetted and another circular incision was performed along the medial site of the original left small uterine horn. Both sides were approximated by 2-0 Dexon using interrupted sutures, without suturing the endometrial lining. Good hemostasis was obtained. In view of the unhealthy appearance of the dilated right fallopian tube, a decision was made to perform a right salpingectomy and histopathology confirmed hematosalpinx. Transfundal 100 mL methylene blue was injected, which passed smoothly to the introitus and freely spilled through the left fallopian tube with no

leak at the suture site. The abdomen was subsequently closed in layers. The patient recovered fully and was discharged three days later.

Follow-up of the patient over a period of 18 months showed that she was had a regular menstrual cycle with good flow for 5 days and minimal dysmenorrhea. A follow-up ultrasound study showed a patent connection between the two horns and an arcuate appearance of the uterus with normal endometrial thickness.

Discussion

Müllerian duct anomalies (MDA) represent a variety of congenital anomalies resulting from arrested development, abnormal formation, or incomplete fusion of the mesonephric ducts. The American Fertility Society separates Müllerian anomalies into seven classes (Table 1). The classification system is useful in most cases, but some cases do not fit into any of these classes. These anomalies have been related to higher rates of infertility, recurrent pregnancy loss, prematurity and other obstetric complications, although many patients may remain asymptomatic. The true incidence of these conditions remains elusive.

Table 1. American Fertility Society classification of Müllerian anomalies.

Class	Description
I	Agnesis
II	Unicornuate
III	Didelphys
IV	Bicornuate
V	Septate
VI	Arcuate
VII	DES-related

The etiology of these developmental anomalies is probably multifactorial, although a genetic component may be present, since a slightly higher frequency is noted in first degree relatives.^{1,2} A rudimentary uterine horn is an extremely rare congenital uterine anomaly. It consists of a normal appearing uterus on one side with a contralateral "horn" or knob. According to the American Society for Reproductive Medicine (ASRM), the classification of rudimentary horns is based on the degree of failure as follows: isolated, with rudimentary horn (communicating), with rudimentary horn (non-communicating), and with rudimentary horn (non-communicating with

no cavity)³⁻⁶ The majority of rudimentary uterine horns do not communicate with the hemiuterus, cervix or vagina, but non-communicating cavitated rudimentary horns are the most clinically significant. They are most likely associated with dysmenorrhea, dyspareunia, infertility and pelvic pain secondary to pelvic adhesions and pelvic endometriosis.

Over 50% of patients with Müllerian duct anomalies will have congenital urinary tract abnormalities. Renal agenesis is more commonly seen in uterus didelphys than in other types of Müllerian anomalies.⁷ Renal agenesis in patients with rudimentary horn is often seen on the ipsilateral site of the rudimentary horn. Hysterosalpingography (HSG) has been the primary diagnostic tool for evaluating uterine cavity abnormalities. However, it cannot detect the presence of a non-communicating horn and cannot be carried out in single girls with an intact hymen. Other methods used to diagnose these condition include three-dimensional ultrasonography (3D), which provides detailed images of both the uterine cavity configuration and adnexal anatomy, especially when image reconstruction is performed.⁸⁻⁹ Magnetic resonance imaging is superior to computerized tomography (CT) and ultrasound in the delineation of congenital anomalies and tumors. Magnetic resonance imaging has a number of advantages, being multi-planar with more optimal tissue contrast and tissue characterization, no radiation hazards and a limited need for intravenous contrast material.⁷ Magnetic resonance imaging should be used for diagnosis rather than for screening if physical examination, hysterosalpingography or ultrasonography suggests the presence of Müllerian anomalies. Magnetic resonance imaging has been suggested as a valuable alternative to laparoscopy and hysterosalpingography for the assessment of MDA.⁹ Minto et al¹³ in a study conducted to evaluate the accuracy of MRI in assessment of adolescent patients with complex Müllerian anomalies, concluded that there was a good correlation between MRI and operative findings in all their cases. They studied 9 patients with Müllerian anomalies, and found that MRI findings were essential for appropriate choice of surgical approach, and type of procedure in 4 of their patients.

It is difficult to reach an early diagnosis in all cases of MDA. The complications of unidentified obstructive MDA includes severe disabling dysmenorrhea, endometriosis, hematocolpos, hematometria, pelvic adhesions and pelvic abdominal mass. Many studies have confirmed the accuracy of MRI as a sensitive and specific diagnostic investigation for MDA.

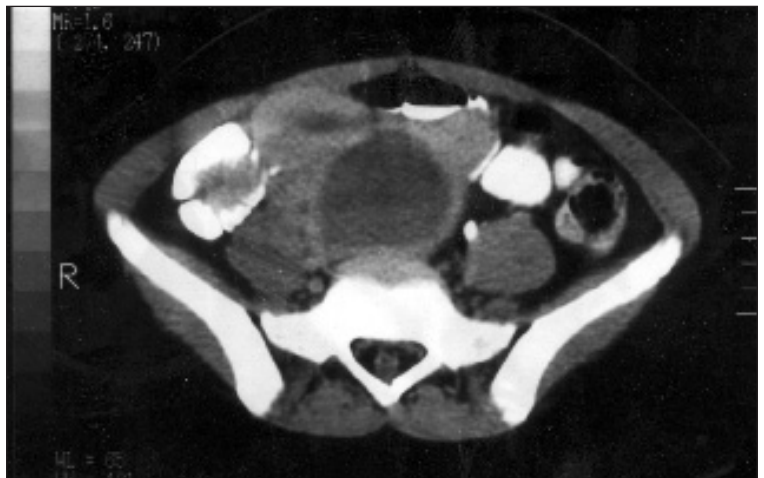


Figure 1. Enhanced CT scan of the pelvis showing large cystic structure.

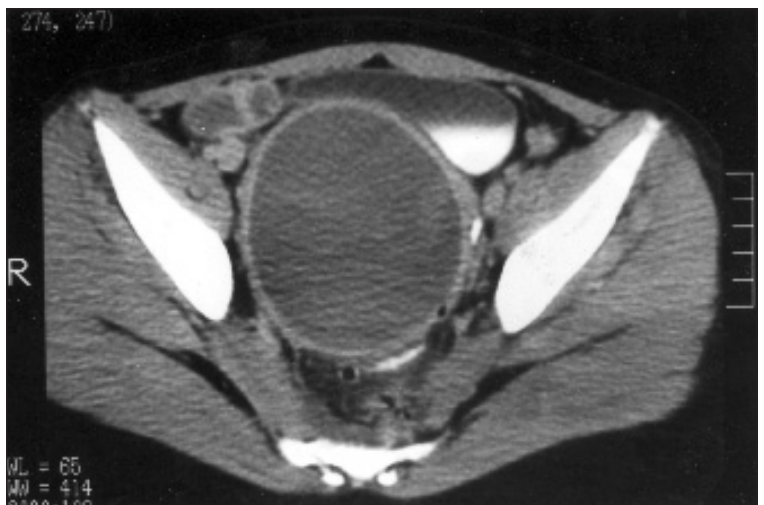


Figure 2. Enhanced CT scan of the pelvis showing large cystic structure.

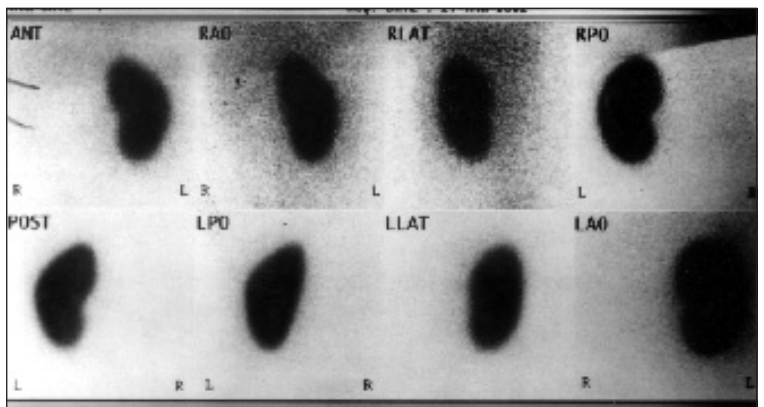


Figure 3. A DMSA isotope scan showing a left kidney of normal size, shape, and position.

Moreover, MRI should now replace hysterosalpingography and diagnostic laparoscopy as second line investigations. The majority of non-communicating rudimentary horns are managed surgically either through laparotomy or laparoscopy for excision of the non-communicating rudimentary horn.^{14, 20} In view of the fact that our young patient was single with very scanty but regular periods, we preferred not to proceed with uterine horn excision but rather to communicate the large non-communicating horn with the small horn, which is in continuity with

the cervix and vagina. To our knowledge this is the first time to report conservative surgery to preserve a non-communicating uterine horn. Unfortunately the obstetric outcome in patients with Müllerian anomalies is not optimal if pregnancy took place before the diagnosis. This carries a risk of early pregnancy wastage, ectopic pregnancy, rupture if located in non-communicating horn, intra-uterine growth restriction and premature labor.²¹⁻²³ In conclusion, Müllerian anomalies vary in presentation and methods of management.

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