Uterine didelphus with obstructed hemivagina

Malak M. Al-Hakeem, MD, ABOG, Samir A. Ghourab, MD, FRCOG, Mohammed R. Gohar, MD, FRCOG, Tariq Y. Khashoggi, MD, ABOG.

ABSTRACT

Complex congenital anomalies of the mullerian ducts can occur in isolation or in association with other developmental disorders. They result from non-development or non-fusion of the mullerian ducts or the failure of reabsorption of the uterine septum. Early diagnosis is necessary to relieve symptoms, optimize preservation of the genital organs and prevent the development of endometriosis. We present a case report to highlight this phenomenon.

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Uterus didelphus is one of the anomalies that may occur as a result of Mullerian duct non-fusion. When complete, the patient has 2 uteri, 2 cervices but the vagina may be single or double. Its incomplete forms may present with varying symptoms and signs. When vaginal septum is incomplete, menstrual efflux may be blocked either at the introitus or at any level up to the cervix. We present a case whose main characteristic is a transverse vaginal septum, which caused most of the presenting symptoms.

Case Report. A 15-year-old girl presented to King Khalid University Hospital, Riyadh, Kingdom of Saudi Arabia, complaining of intermittent left loin pain of 10-months duration with increased severity during menstruation. She reached menarche at 13-years-old. She experienced severe disabling dysmenorrhea, whose intensity increased progressively with each cycle. Her menstrual cycles were regular, bleeding for 7 days out of 28 on examination, she was found to be a well developed young lady. Her secondary sexual characteristic was normally formed. There were no abnormalities in her respiratory and cardio vascular systems, abdominal examination did not reveal any palpable masses, inspection of the introitus did not reveal any bulge as one would expect in most cases of hematocolpus this turned out to be due to the fact that the obstructing septum was transverse and high in the left lateral aspect of the vagina. Ultrasound scan was performed, this suggested the presence of bicornuate uterus with a hematocolpus in the left cornua. Later, magnetic resonance imaging (MRI) was performed with (T1 and T2 WI obtained in the sagittal, coronal T1 fat-saturated post Gadolinium) and this showed double uterus with 2 cervices and duplication of the vagina. It also revealed left hematocolpus. Intravenous pyelogram revealed normal right kidney and absent left kidney. With collection of the history, physical examination and radiological investigations, a provisional diagnosis of uterus didelphus with 2 cervices and 2 vaginas was made, we felt that the

From the Department of Obstetrics & Gynecology, King Khalid University Hospital, Riyadh, *Kingdom of Saudi Arabia.*

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Address correspondence and reprint request to: Dr. Malak M. Al-Hakeem, Assistant Professor and Consultant, Department of Obstetrics & Gynecology, King Khalid University Hospital, PO Box 8413, Riyadh 11662, *Kingdom of Saudi Arabia.* Tel. +966 (1) 4670818. Fax. +966 (1) 4679347. E-mail: kmmtmm@hotmail.com

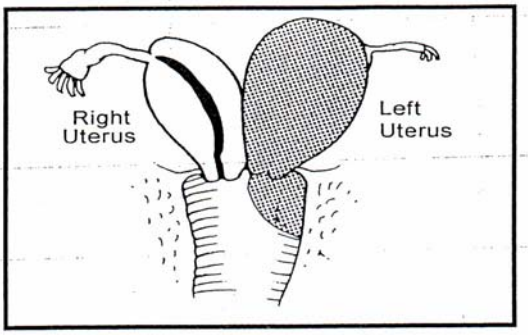

Figure 1 - Laparotomy revealed uterus didelphus, with left side hematocolpus and normal right uterus and tube.

presence of the hematocolpus coupled with normal menstruation meant that one of the vaginas was obstructed. These findings were explained to the patients and her parents. They were informed that it would be necessary to remove the obstructed uterus, which was the cause of her pain. Permission for the surgery was granted. At laparotomy, the right uterus and tube were normal. The left uterus was distended with blood. The left tube originated from the apex of the left uterus and it looked rudimentary (**Figure 1**). On injection of Mehtylene blue dye, spillage was seen from the right side only. No endometriosis was seen. Excision of the left uterus and the left tube was carried out. The blood collected in the vagina was drained out. The patient has been followed up as an out-patient for 18 months now and she maintains that her cycles remain regular and her dysmenorrhea has improved.

Discussion. Mullerian duct anomalies (MDA's) are relatively common, occurring in one in 200-600 women of child-bearing age and result from defect in fusion, migration or lack of recanalization.[1] The uterus is the most frequently affected site, with resulting structural abnormalities ranging from septations to the presence of double uterus and cervices as in the didelphic uterus. The etiology of these developmental anomalies is probably multifactorial, although a genetic component may be present since a slight higher frequency is noted in first-degree relatives.[2,3] Over 50% of patients with Mullerian duct anomalies will have congenital urinary tract abnormalities.[1-4] Renal agenesis is more commonly seen in uterus didelphus than in other types of MDA's. Renal agenesis in patients with uterus didelphus is often seen on the ipsilateral site of an obstructing, transverse, or hemivaginal septum.[4,5] Traditionally hysterosalpingography (HSG) has been the primary diagnostic tool used to evaluate uterine cavity abnormalities.[6] However, it cannot detect the presence of non-communicating horn and it cannot be carried out in single girls with intact hymen. Other methods used to diagnose MDA's include zoography, laparoscopy, hysteroscopy and magnetic resonance imaging (MRI). Three-dimensional (3D) ultrasonography offers several advantages over conventional 2 dimensional (2D) scanning. Three-dimension (3D) sonography provides detailed images of both uterine cavity configuration and adnexal anatomy, especially when image reconstruction is performed.[6] Three dimensional (3D) ultrasound was not carried out in our patient, as it was not available in the hospital at that time. Magnetic resonance imaging is a very useful tool in the diagnosis of MDA and the presence of any renal anomalies. It provides detailed information without exposing the patient to ionizing radiation as in HSG or to invasive technique requiring anesthesia, as in laparoscopy. Magnetic resonance imaging is superior to computerized tomography (CT) and ultrasound in the delineation of congenital anomalies and tumors. Magnetic resonance imaging has advantages of being multi-planar with more optimal tissue contrast and tissue characteristics, no radiation hazard and limited need for intravenous contrast material.[7] Magnetic resonance imaging should be used for diagnosis rather than for screening if physical examination, hysterosalpingogram or sonogram suggests the presence of Mullerian anomalies. Magnetic resonance imaging has been suggested as a valuable alternative to laparoscopy and hysterosalpingography for the assessment of MDA.[8,9] Minto et al[10] in a study to evaluate the accuracy of MRI in assessment of adolescent patients with complex Mullerian anomalies concluded that there was a good correlation of MRI and operative findings in all their cases, when they studied 9 patients with Mullerian anomalies, they found that MRI findings were essential for appropriate choice of the surgical approach, and type of procedure in 4 of their patients. The complications of unidentified obstructive MDA are hematocolpos and hematometra. Early diagnosis of a didelphus uterus and surgical removal of non-communicating uterus is justified, in the presence of disabling dysmenorrhea and also as of the likely development of endometriosis and pelvic adhesions, due to retrograde expulsion of menstrual products by the fallopian tube in the non communicating cavitary uterus. Many studies have confirmed the accuracy of MRI as a sensitive and specific diagnostic investigation for MDA. It is likely that ultrasound will remain the initial investigation for all patients with MDA. However, MRI should now replace hysterosalpingogram and diagnostic laparoscopy as 2nd line investigations. The conventional operation for uterus didelphus with hematometria in a one sided non-communicating uterus consists of excision of that non communicating uterus. Such patients have increasing dysmenorrhea beginning soon after the

menarche. The diagnosis is often missed as the patient has cyclic vaginal bleeding accompanied by the dysmenorrhea.

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