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A case of complete non-fusion of Müllerian ducts associated with Bladder exstrophy: A case report

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Abstract:
Uterine didelphys is a relatively rare congenital anomaly of the mullerian ducts. However, its association with bladder exstrophy, though embryologically well understood, is very rare. Here, we present such a case of uterus didelphys with atretic right lower hemivagina with a history of bladder exstrophy. She came to us unable to perform sexual intercourse.
Keywords: *Bladder exstrophy, uterine didelphys, mullerian anomaly, case report*

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I. Introduction:

Development of the female genital tract is a complex process that depends on a series of events involving cellular differentiation, migration, fusion and canalization.

The mullerian ducts form fallopian tubes, uterus and proximal two-thirds of the vagina, while urogenital sinus forms the distal one-third of vagina and the urethra. Failure in any of the above processes will result in congenital anomalies of genital tract. The association of failure of fusion of the mullerian ducts with other genitourinary anomalies is well known. Uterine didelphys is an abnormality of lateral fusion of the ducts resulting in two separate uteri. According to the American Fertility Society, uterine didelphys will fall into Class III. The incidence is 5 to 7% of all uterine anomalies.(1) Kidneys are derived from the mesonephros, which are closely related to the Mullerian ducts and hence, Mullerian anomalies can be associated with problems of the urinary tract. However, bladder exstrophy is an uncommon association with uterine didelphys and only a few have been reported in the past.(2)(3)

Bladder exstrophy is a congenital anomaly of the urinary tract whose feature is the exposure of the posterior wall of the bladder to the exterior. It occurs in one of 50,000 live births. It is inherited in a multifactorial fashion. In women, considerable disruption of the external genitalia may be associated with bladder exstrophy.(3)

Here, we present such a case of uterus didelphys with atretic right lower hemivagina with a history of surgery for correction of bladder exstrophy.

Presentation of the case:

A married 21 year old woman presented complaining of inability to perform sexual intercourse and a history of bladder surgery. She underwent reconstructive surgery in early infancy for bladder exstrophy and another surgery later in her childhood to remove a large bladder stone. She also has stress urinary incontinence.

On examination, a thick vertical hyperpigmented infraumbilical scar was noted. The symphysis pubis was not palpable. The clitoris was absent and two pinhole vaginal openings were noted beneath the external urethral meatus.

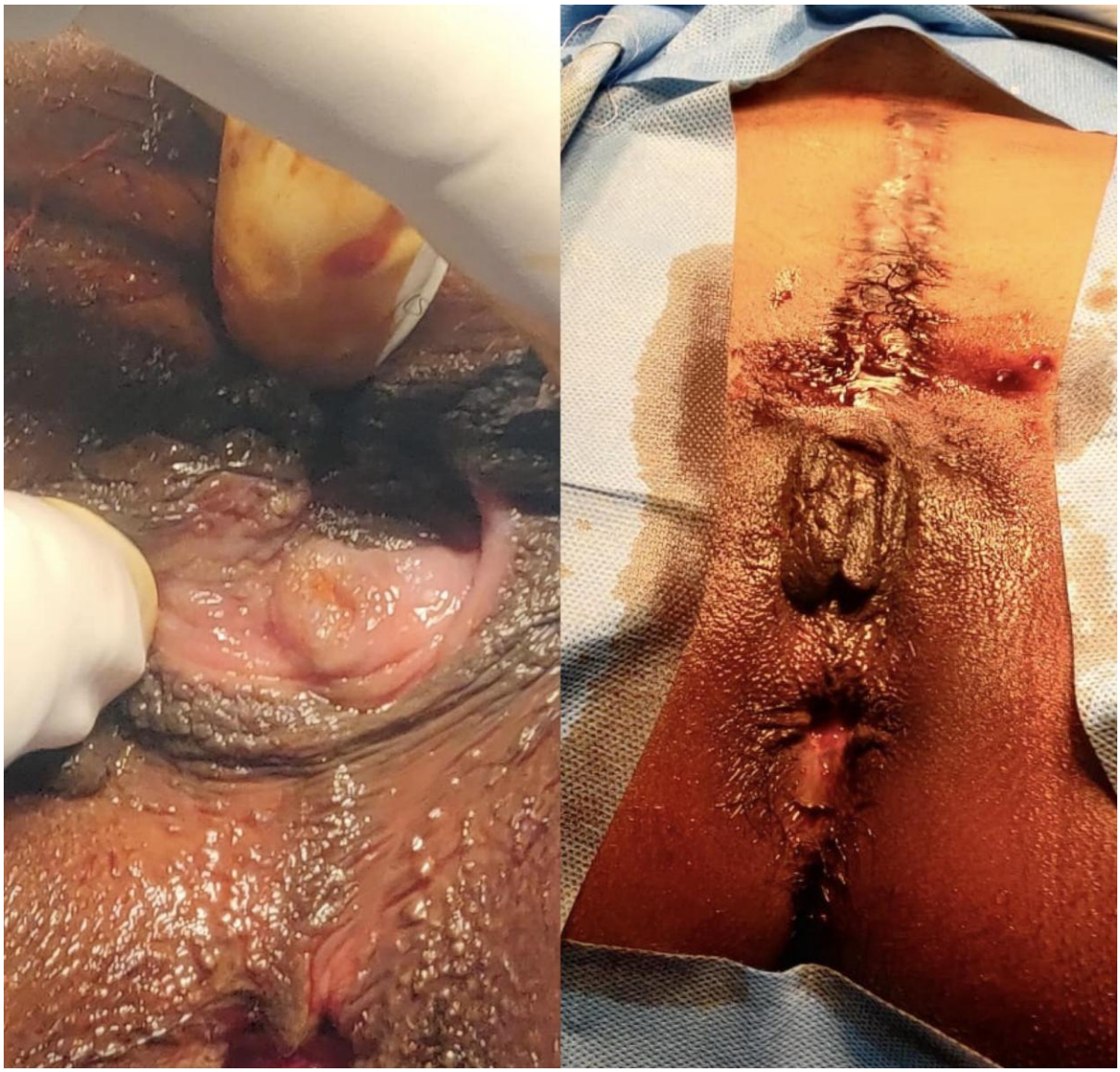

Fig. 1 Right subset of the image shows the vertical scar of previous surgery for bladder exstrophy repair and also shows poorly formed external genitalia. The left subset shows pinpoint introitus which upon hysteroscopy revealed adequate normal vagina except a total vaginal septum with bicollis.

Ultrasound of the abdomen and pelvis could only pick up bilateral bulky ovaries with a polycystic pattern. This was followed up with an MRI pelvis which revealed two independent uteri with wide-spaced divergent apices showing normal endo-myometrial zonal anatomy and complete duplication of the cervix and upper third of vagina. The left vagina appears normal whereas the right hemivagina is hypoplastic or atretic. In addition, MRI also shows widely separated pubic symphysis with midline anterior pelvic wall surgical scar extending into the deep facial planes up to the antero-inferior wall of partially distended urinary bladder. Kidneys were normal in number, location and size.

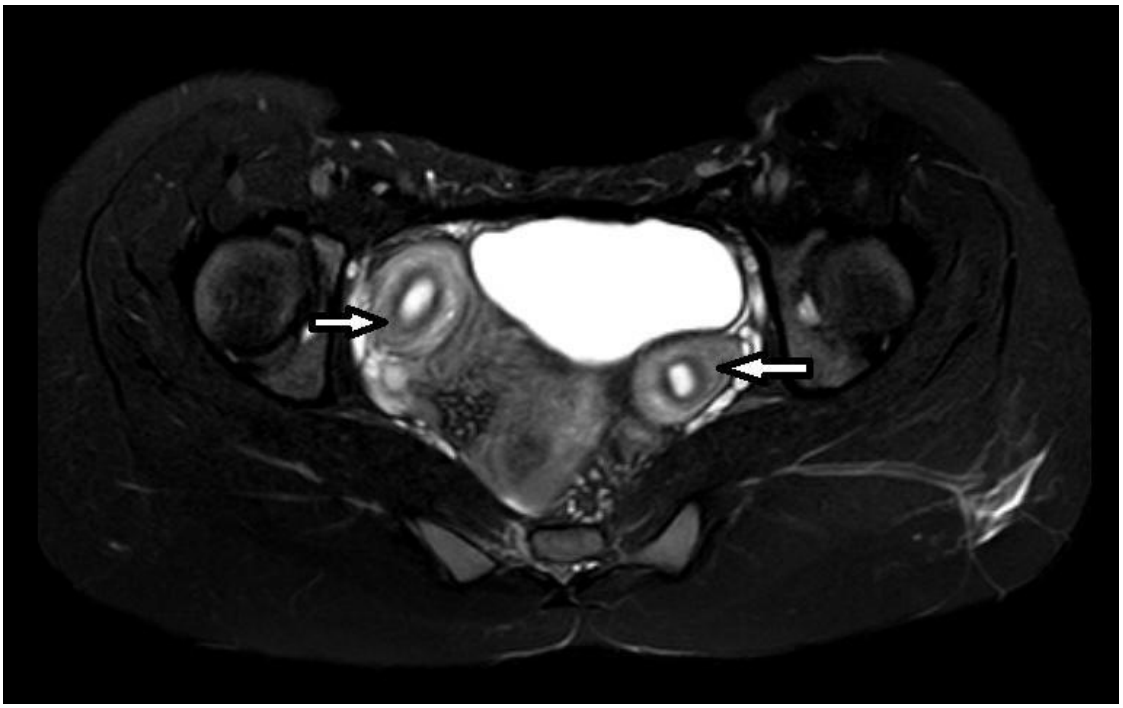

Fig. 2 Magnetic Resonance Imaging showing two independent divergent uterine horns with an abnormally low placed bladder between the two horns.

Henceforth, the diagnosis of uterus didelphys with atretic right lower hemivagina and mild bilateral polycystic ovarian syndrome with a history of surgical repair for bladder exstrophy was made.

Under spinal anaesthesia, vaginoscopy was performed through the two distinct pinhole openings. Two separate vaginal canals were visualized with a thick vaginal septum and both canals ended with separate cervices, each consisting of a cervical canal. The vaginal septum was identified and resected. A week later, serial vaginal dilatations were done. The patient was counselled and educated about self dilatation which she followed on a daily basis at home. This was monitored on a weekly basis in the outpatient department. Two months later the patient was encouraged to have an active sexual life.

II. Discussion:

We presented a case of a 21 year old woman who has uterine didelphys and septate vagina with a history of surgical correction of congenital bladder exstrophy. Congenital pelvic anomalies are often complex and frequently occur as disorders of both the urinary and reproductive tracts due to the pattern of embryological development.[4]

Congenital uterine anomalies represent a spectrum of morphological abnormalities from varying degrees of non-fusion of Mullerian ducts occurring in 1-5% of the general population. Its association with bladder exstrophy is extremely rare. American Fertility Society (AFS) classifies congenital uterine anomalies. Uterus didelphys is the most rare one with an incidence of 5-7% of all Mullerian duct anomalies.[1] 75% of uterine didelphys are associated with longitudinal vaginal septum and an occasional transverse vaginal septum.[3]

The mullerian ducts appear during the first week of gestation. They grow caudally and eventually contact the posterior wall of the urogenital sinus at the mullerian tubercle. The ducts then fuse and canalize to form the fallopian tubes, uterus, and upper two thirds of the vagina eventually arise. Uterine anomalies occur due to varying degrees of failure in the above described process. Uterus didelphys results from complete lack of fusion. Vaginal hypoplasia occurs when the distal ends of the mullerian ducts fail to canalize.[3] Bladder exstrophy occurs secondary to rupture of an abnormally persistent cloacal membrane after the completion of the urorectal septum. This interferes with the fusion of the müllerian ducts which leads to associated urogenital anomalies.[3] Such association is rare, and only a handful of cases of combined uterus didelphys, septate vagina, and bladder exstrophy have previously been reported.[3][4]

In patients with urinary tract anomalies, it is mandatory to evaluate the reproductive tract to rule out any coexisting Mullerian duct anomalies. Radiographic imaging can be utilized to identify and classify co-existing anomalies of the reproductive tract. Ultrasound and hysterosalpingography are used for screening but both modalities have their limitations. In our case, ultrasonography could not pick up on the uterine anomaly, which is diagnostic in 50% of patients and a transvaginal ultrasonography, which is diagnostic in 92% of

patients, was not possible due to the small vaginal orifices.[2] When an associated genital tract anomaly is not picked up with ultrasonography, it should further be evaluated with an magnetic resonance imaging. An MRI is accurate with 100% precision in detecting genital tract anomalies. It is the gold standard for detecting and classifying Müllerian anomalies.[2][3]

The goal with treatment was to achieve normal sexual function as intercourse was limited by the narrow introitus. A vaginoplasty was performed which created a single larger vaginal orifice but required serial vaginal dilators over a period of time to ensure the introitus was large enough for normal sexual function. Though, 40% of pregnancies in women with uterine didelphys end in spontaneous abortions, a normal endo-myometrial zonal anatomy revealed in the MRI suggested a good prognosis for conceiving if the patient wished for it. Corrective surgery like metroplasty is required only with recurrent miscarriages.

III. Conclusion:

Uterine didelphys is a rare Mullerian duct anomaly and its association with bladder exstrophy makes this case unique. Congenital anomalies of the urinary tract and the lower genital tract should be followed up with an MRI for accurate diagnosis. Didelphys uterus with a normal endo-myometrial junction has a better reproductive outcome as compared to other mullerian anomalies. Corrective reconstructive surgery for uterine didelphys is required only when normal functioning is not possible without it.

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