JBR–BTR, 2011, 94: 16-18.

UTERUS DIDELPHYS WITH OBSTRUCTED HEMIVAGINA AND RENAL AGENESIS: MRI FINDINGS

A. Talebian Yazdi, K. De Smet, C. Ernst, B. Desprechins, J. de Mey[1]

Müllerian duct abnormalities (MDA) are developmental disorders leading to dysmorphism of the female genital tract. Currently the Buttram and Gibbons classification of these entities is widely used. We present a case of a young girl with uterus didelphys and ipsilateral renal agenesis.

Key-word: Uterus, abnormalities.

Müllerian duct abnormalities (MDA) are developmental disorders leading to dysmorphism of the female genital tract. The disruption of the normal embryologic fusion of the paramesonephric (Müllerian) ducts or the non-resorption of the uterine septum is believed to be the crucial element in this entity (1, 2). Buttram and Gibbons have created a widely used classification of MDA (3)

MDA are rare syndromes and scarcely reported on in English literature. But the introduction of the modern MRI equipment has led to improved recognition and familiarity with these complex malformations.

Clinically the patients present with nonspecific symptoms around the time of menarche. Imaging can be helpful in the correct diagnosis, preventing delayed surgical intervention and possible infertility.

In this report we present a case of uterus didelphys with an obstructed hemivagina and ipsilateral renal agenesis (Buttram and Gibbons class III) and a brief review of literature.

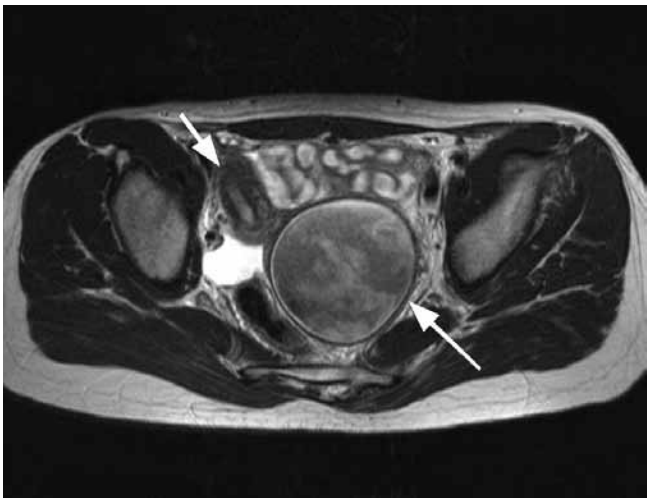


Fig. 1. — Axial T2WI demonstrating the presence of a didelphic uterus with a small right (short arrow) horn and an abnormally distended left horn (large arrow) due to a fluid collection consistent with old menstrual blood.

Case report

A 12-year-old female patient presented at our emergency department with intermittent abdominal pain and heavy vaginal blood loss. The menstrual cycle had started 6 months before and was inconspicuous until the last 2 months. At that time menstruation was accompanied by intense pains and heavy blood loss.

Initial clinical workup did not lead to a specific diagnosis. Pelvic ultrasound was performed and showed hematometra with suspicion of uterine dysmorphism. The patient was scheduled for a MRI of the pelvis.

The MRI protocol consisted of standard T1 and T2 sequences with and without fat suppression and reconstructions in the three anatomical planes. A total duplication of the uterus was demonstrated. The vagina was divided into separate chambers by a septum. The left hemivagina and hemi-uterus were distended and contained a fluid collection. The signal characteristics of this collection were compatible with old (menstrual) blood (Fig. 1-4).

The right hemi-uterus had a normal appearance; the right hemivagina was displaced by the fluid containing left hemivagina. The ovaries had a normal appearance.

Ipsilaterally to the affected side of the internal genitalia, absence of the left kidney was noted (Fig. 5).

The MRI results were consistent with uterus didelphys with an obstructed hemivagina and ipsilateral renal agenesis.

The patient underwent gynaecologic intervention, the obstructed hemivagina was opened and the septum between the two vaginas was excised. There were no major complications and the next day the patient left our hospital in good condition.

Discussion

MDA are developmental disorders that can lead to a variety of abnormalities in the female urogenital tract. The reported incidence in English literature is 0.5-5% (1, 2). Buttram and Gibbons have classified the various MDA into 6 categories (3).

The presented case illustrates the imaging findings of uterus didelphys with an imperforate hemivagina and

From: 1. Department of Radiology, University Hospital Brussels, UZ Brussel, Brussels, Belgium
Address for correspondence: Dr. A. Talebian Yazdi, MD, Department of Radiology, University Hospital Brussels, UZ Brussel, Laarbeeklaan 101, B-1090 Brussel, Belgium
E-mail: atalebiany@hotmail.com

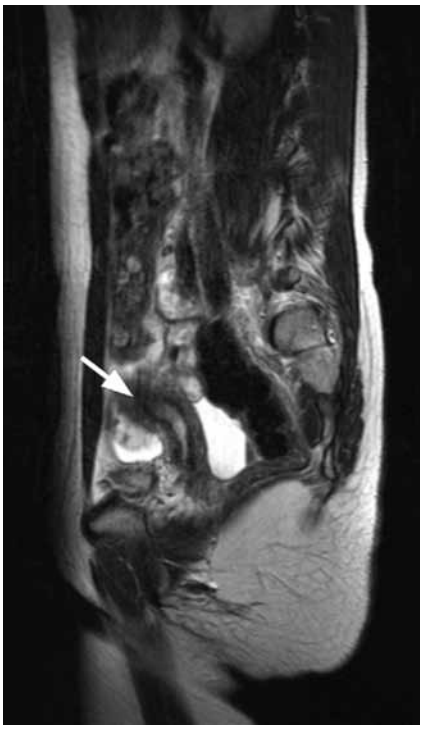


Fig. 2. — Right parasagittal T2WI demonstrating the small right horn of the didelphic uterus (arrow).

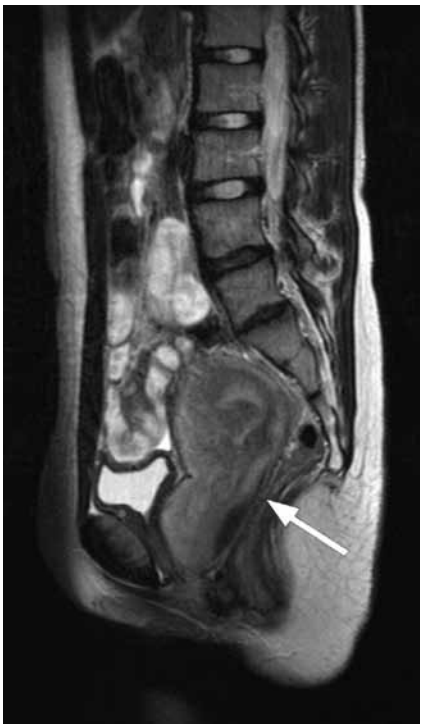
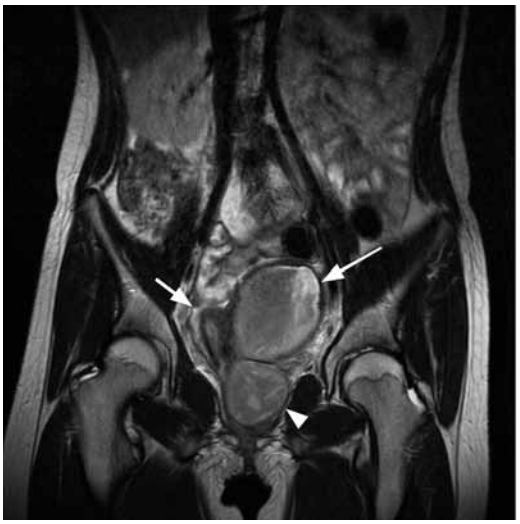


Fig. 3. — Left parasagittal T2WI demonstrating the abnormally distended left horn of the didelphic uterus (arrow) due to a fluid collection consistent with old menstrual blood.



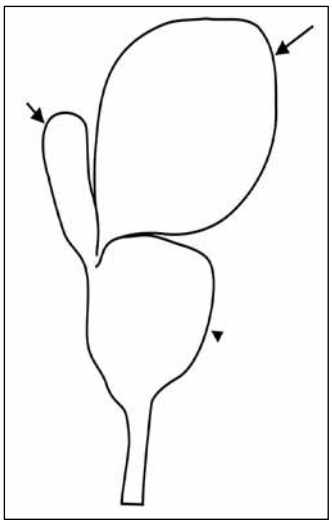


Fig. 4. — Coronal T2WI and drawing, demonstrating the morphology of the didelphic uterus with the small right (short arrow) horn and the abnormally distended left horn (large arrow). The vagina (arrowhead) is also abnormally distended due to the persistent vaginal septum.

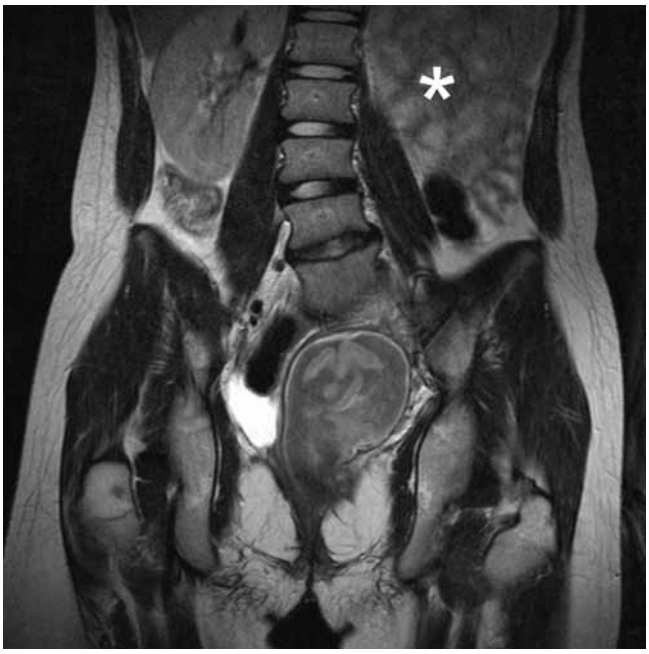


Fig. 5. — Coronal T2WI demonstrating the absence of the left kidney in the left renal fossa (asterisk).

an absent left kidney (Buttram and Gibbons class III anomaly). The presence of two separate and divergent uterine horns, two cervices and a duplicated vagina with partial obstruction are characteristic for this entity. Knowledge of the normal embryologic events aids in the understanding of this complex dimorphic syndrome. The cascade of events in the development of the female genital tract occurs during the 6th and 22nd week of gestational age. The paramesonephric (Müllerian) ducts move toward the midline, fuse and insert in the urogenital sinus. Any disruption in this cascade of events leads to abnormalities of the genital tract. In the specific case of uterus didelphys there is a complete nonfusion of the Müllerian ducts leading to the duplication, of the uterus and the vagina. Disorders of the urologic tract, in our case the absence of the left kidney, could be explained by the close

embryologic relationship between the mesonephric (Wolffian) ducts and the paramesonephric ducts. Various teratologic influences are believed to cause concomitant urologic and gynaecologic malformations (4-6)

MRI, with its superior soft tissue resolution, often leads to a straightforward diagnosis of complex Müllerian duct abnormalities. Therefore it is considered the best non-invasive tool in the work-up of malformations of the urogenital tract.

Knowledge of the main imaging findings of uterus didelphys and other MDA will aid in the timely and correct diagnosis. Complications of delayed diagnosis are infection with subsequent abscess formation and hysterectomy with or without oophorectomy leading to infertility.

Conclusion

Müllerian duct abnormalities encompass a variety of dimorphic changes in the female genital tract, often with associated urologic anomalies. In this paper the MR characteristics of uterus didelphys with an imperforate hemivagina and an absent left kidney were discussed. Familiarity of the radiologist with the imaging findings of this rare syndrome is important, because delayed diagnosis increases the chance of complication and possible infertility.

References

1. Nahum G.G. Uterine anomalies: how common are they and what is their distribution among subtypes? *J Reprod Med*, 1998, 43: 877–887.
2. Stampe Sorensen S. Estimated prevalence of müllerian anomalies. *Acta Obstet Gynecol Scand,* 1988, 67: 441-445.
3. Buttram V.C. Jr, Gibbons W.E. Müllerian anomalies: a proposed classification (an analysis of 144 cases). *Fertil Steril*, 1979, 32: 40-46.
4. Anderson K.A., McAninch J.W. Uterus didelphia with left hematocolpos and ipsilateral renal agenesis. *J Urol*, 1982, 127: 550-553.
5. Yoder I.C., Pfister R.C. Unilateral hematocolpos and ipsilateral renal agenesis: report of two cases and review of the literature. *AJR*, 1976, 127: 303-308.
6. Eisenberg E., Farber M., Mitchell Jr, G.W. Complete duplication of the uterus and cervix with a unilaterally imperforate vagina. *Obstet Gynecol* 1982, 60: 259-262.