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Case Report

Mullerian ducts anomaly of 2 divergent uterine horns with cervicovaginal hypoplasia: Is it considered unclassified or under the spectrum of Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome?☆

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ABSTRACT

The Mullerian ducts give rise to the upper part of the female reproductive system, including the uterus, cervix, upper two-thirds of the vagina, and fallopian tubes, which undergo specific processes of development, fusion, and resorption. Any failure in this process will lead to Mullerian duct anomaly (MDA).

We present a unique and complex case of MDA, signifying the wide variability and simultaneous existence of combined abnormalities in 1 patient, which do not always fit under a single or particular class from the known classification systems. Therefore, subclassifications may be necessary for each part alone (uterus, cervix, and vagina) or incorporating more than 1 class for a single case. It also shows the role of imaging in the diagnosis; considering that magnetic resonance imaging (MRI) is the standard modality for a detailed description of the reproductive system and its anomalies.

Introduction

Mullerian duct anomaly (MDA) is a unique entity with a widely variable and complex spectrum of abnormalities. Its prevalence varies substantially according to the population under study, with mean prevalence in the general population up to 7% [1,2] and 25% in women with a history of infertility and miscarriage [1].

There are multiple classification systems in the literature. The most widely used system was published in 1988 by the American Society for Reproductive Medicine (ASRM); it includes seven classes [3] and was updated in 2016 [1]. The other was founded by the European Society of Human Reproduc-

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tion and Embryology and the European Society for Gynaecologic Endoscopy (ESHRE/ESGE) in 2013, which is anatomy-based, classifying the anomalies according to the morphology of the uterus, cervix, and vagina, independently [3].

Knowledge of the embryology and formation mechanism of the female reproductive system is essential in understanding the pathophysiology of MDA. It originates from different structures; the paired Mullerian ducts give rise to the uterus, fallopian tubes, cervix, and upper two-thirds of the vagina [4], which undergo specific processes of development, fusion, and resorption. Any failure in the process will lead to MDA. The ovaries and distal one-third of the vagina originate from the primitive yolk sac and sinovaginal bud, respectively [4]. Therefore, MDA is not associated with anomalies of the external genitalia or ovarian development [4]. It can be isolated or associated with different anomalies, especially renal ones.

Patients often present with primary amenorrhea, infertility, obstetric complications, and endometriosis [4]. They may also present with obstructive symptoms such as hematometra, hematocolpos, and cyclic lower abdominal pain or may be asymptomatic, depending on the functional status of the endometrium [2].

Imaging has a vital role in the detection, diagnosis, and complete characterization of MDA. Transabdominal and endovaginal US are the first-line modalities [1]. The MRI is the imaging standard of reference because it is noninvasive, does not involve ionizing radiation, has multiplanar capability, allows excellent soft-tissue characterization, and permits a greater interrogation field than ultrasound [4,5]. It enables a detailed anatomical description of the uterus, cervix, vagina, and ovaries [3] in addition to detection of possible associated urological anomalies. MRI has a reported accuracy of up to 100% in evaluating MDA [2]. Hysterosalpingography (HSG) remains the standard modality for evaluating fallopian tube patency in patients with infertility [1] but has a limited role in fully evaluating MDA.

Case report

A 25-year-old woman with a history of primary amenorrhea went to the OB/GYN outpatient clinic in 2017 and had a transabdominal US (Fig. 1) and pelvic MRI, which re-

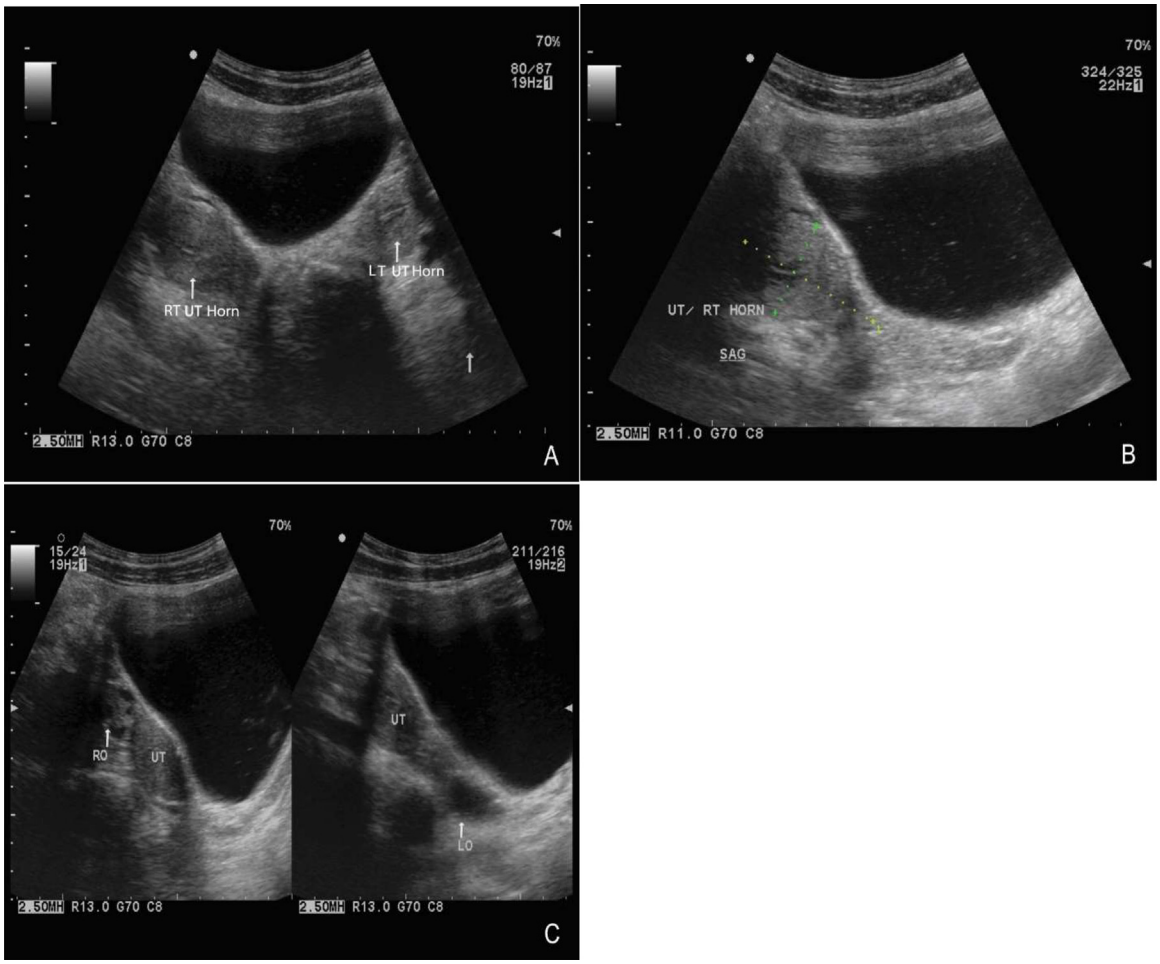

Fig. 1 – Transabdominal ultrasound in 2017 and 2023 done for the patient demonstrating similar findings. (A) show widely separated 2 uterine horns with normal myometrial echogenicity and central thin echogenic endometrial stripe on either side (White arrows). (B) On the longitudinal view, no cervix or upper vagina could be seen. (C) show bilateral normal appearing ovaries containing physiological follicles (White arrows). No abnormal pelvic masses.

vealed MDA. However, she reported 2 episodes of blood spotting/menstruation at the age of menarche, around 13 years old.

Then, she returned to the obstetric clinic in 2022, married for the second time since 2019 with a history of primary infertility and seeking pregnancy.

On examination, the patient is skinny and has normally developed secondary sex characteristics with normal breast tissue, normal hair growth, and marked body acne. There were no palpable abdominal masses. Local and vaginal speculum examination show normal external genitalia, blind vagina, and normal vaginal length with a small cervical dimple on the left posterior wall of the vagina. During the bimanual exam, the uterus was not palpable. The patient has had regular sexual intercourse without any issues and was medically free.

There was no significant past surgical history apart from a diagnostic laparoscopy in the USA in 2019. This laparoscopy revealed no cervix, blind vagina, separated didelphys uterus, hypoplastic/absent cervix, and stage 1 endometriosis. She also had 3 trials of IVF, which failed twice in 2019 through trans-tubal ET and once in 2020 through transvaginal transmyometrium ET.

She has had normal laboratory investigations, including TSH, LH, FSH, estradiol, progesterone, and testosterone levels.

In January 2023, the patient had another pelvic US and MRI (Fig. 2), demonstrating the same findings in 2017 of 2 widely separated noncommunicating uterine horns with preserved normal zonal anatomy and a normally appearing endometrial cavity. There is a rudimentary tissue or band of low signal intensity in T2 and T1 located in the midline converging between the uterine horns and extends along the expected location of the cervix and upper two-thirds of the vagina representing the hypoplastic cervix and upper vagina. The lower one-third of the vagina and ovaries are normal. The kidneys are in their normal location, with no identifiable renal anomalies.

Discussion

Depending on the present classification systems, we proposed that our case can be classified by the ASRM (Table 1 and Fig. 3) into (class Ie), considering the presence of combined cervical and vaginal hypoplasia with no specific class could be assigned for the divergent uterine horns [1,3,4]. By the ESHRE/ESGE (Table 2 and Fig. 4), we classify it as (U5a/b, C4, and V0/4) [1,3].

While reviewing the literature, we found that some published cases had findings like ours. Although the images are identical in some cases, there was wide variability in describing and classifying the anomalies. Mahdavi et al. [6] published the most similar cases, describing them as uterus dysplasia associated with cervicovaginal agenesis, and the patient was

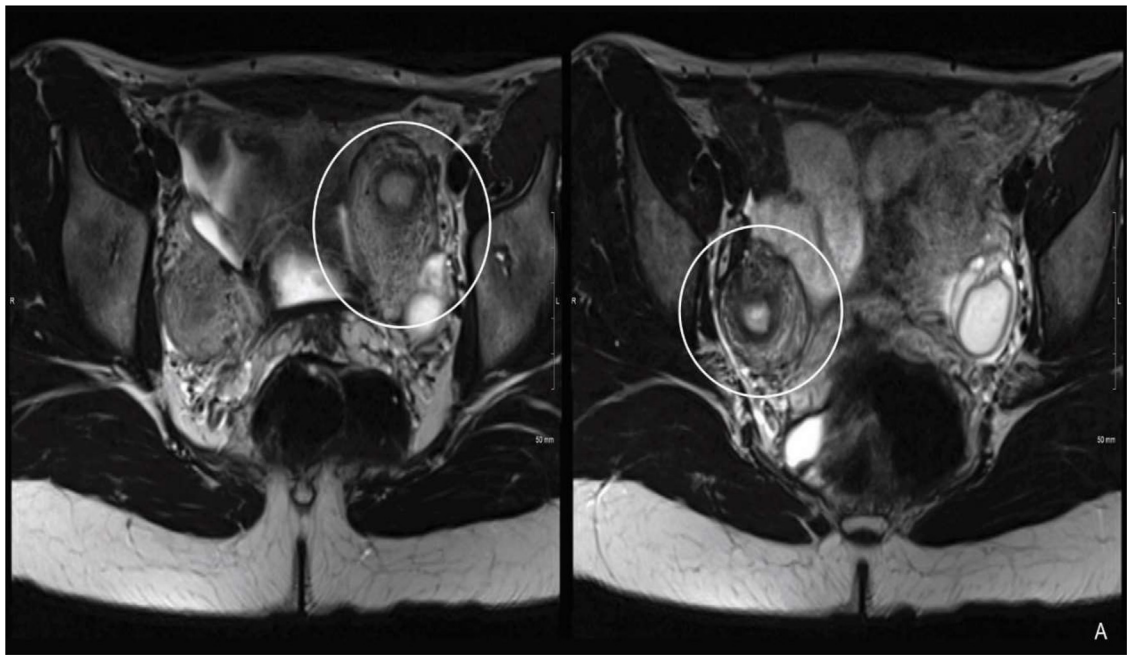

Fig. 2 – Magnetic resonance imaging of the pelvis confirming the ultrasound findings with additional details. Non-fat sat axial T2 weighted images (A, B). (A) show 2 widely separated uterine horns (White circles) with well appreciated normal zonal anatomy and normally appearing non-communicating endometrial cavities of high signal intensity. (B) At lower level there is a dark signal intensity band/tissue at expected location of the lower uterine segment and cervix (Yellow arrow) and normal lower one-third of the vagina (White arrow). (C) Non-fat sat sagittal T2 weighted image shows the poor anatomical differentiation and development of the cervix/upper vagina (Yellow arrow) and the present lower vagina (White arrow). A thin dark T2 signal intensity fibrous band between the 2 horns converging into midline was seen (not shown here). (D) Coronal fat sat T2 weighted image shows bilateral normal ovaries with multiple physiologic follicles. (E) Non-fat sat coronal T2 HASTE shows normal kidneys are present in its anatomical position.

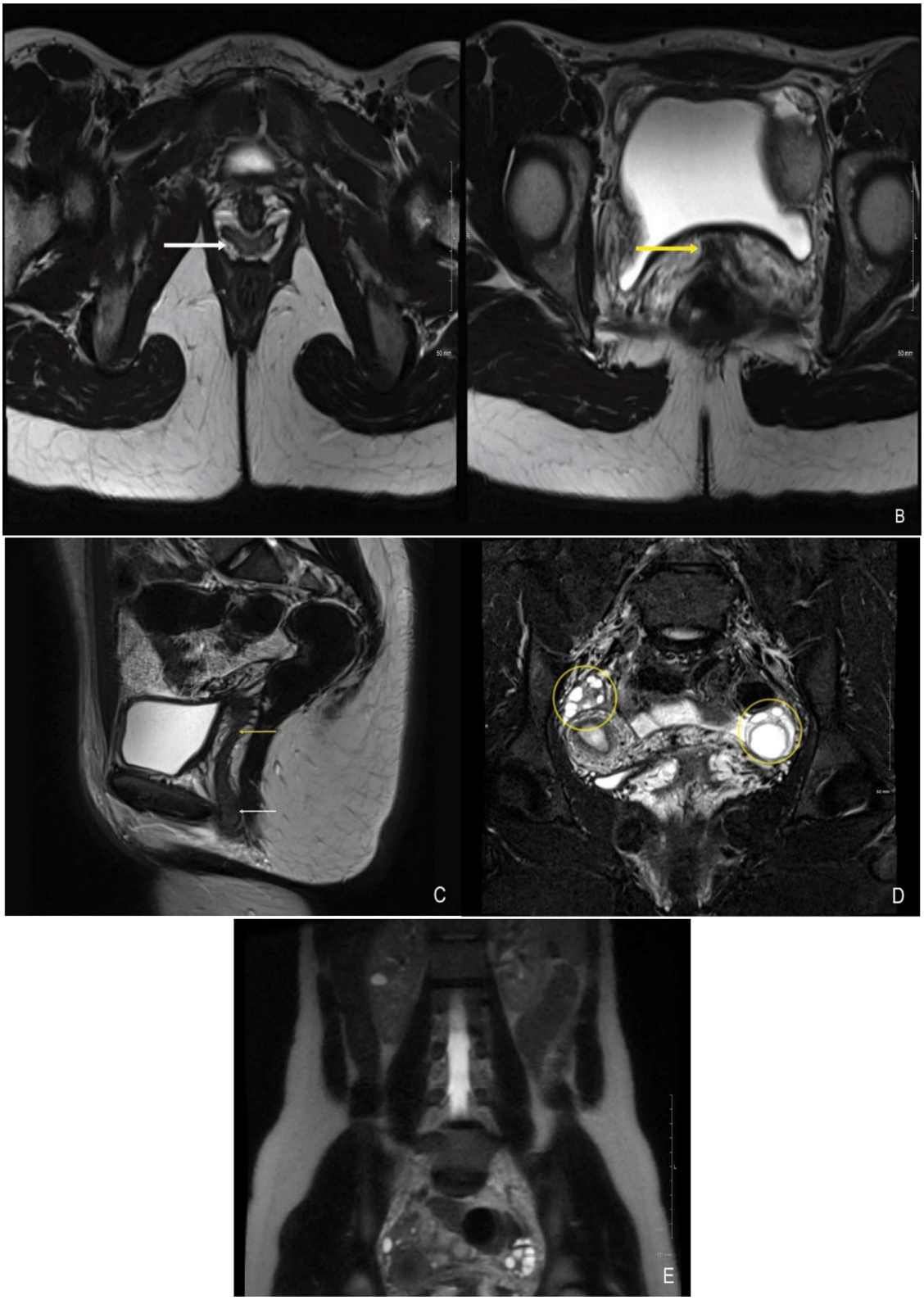

Fig. 2 – Continued

Table 1 – American Society for Reproductive Medicine classification [3].

Class	Classification
Class I	Agenesis or hypoplasia – (a–e) (vaginal or cervical or fundal or tubal or combined)
Class II	Unicornuate – (a–d) (communicating horn or non-communicating horn or no cavity or no horn)
Class III	Uterine didelphys
Class IV	Bicornuate uterus (a and b – Complete or partial)
Class V	Septate uterus (a and b – Complete or partial)
Class VI	Arcuate uterus
Class VII	Diethylstilboestrol related

Table 2 – The European Society of Human Reproduction and Embryology and European Society for Gynecological Endoscopy classification (U, uterine; C, cervical; V, vaginal) [3].

	Uterine anomaly			Cervical/Vaginal anomaly
	Main class	Sub-class		Coexistent class
U0	Normal uterus			
U1	Dysmorphic uterus	U1a - T-shaped uterus	C0	Normal Cervix
		U1b - uterus infantile	C1	Septate
		U1c - others	C2	Double “normal”
U2	Septate uterus	U2a - partial septate uterus	C3	Unilater aplasia or dysplasia
		U2b - complete septate	C4	Aplasia or dysplasia
U3	Bicorporeal uterus	U3a - partial bicorporeal		
		U3b - complete bicorporeal		
		U3c - bicorporeal septate uterus	V0	Normal vagina
U4	Hemi-uterus	U4a - – hemi-uterus with a rudimentary (functional) cavity	V1	Longitudinal nonobstructing vaginal septum
		U4b - – hemi-uterus without a rudimentary (functional) cavity	V2	Longitudinal obstructing vaginal septum
U5	Aplastic uterus	U5a - aplastic uterus with rudimentary (functional) cavity	V3	Transverse vaginal septum or imperforate hymen
		U5b - aplastic uterus without rudimentary (functional) cavity	V4	Vaginal aplasia
U6	Unclassified cases			

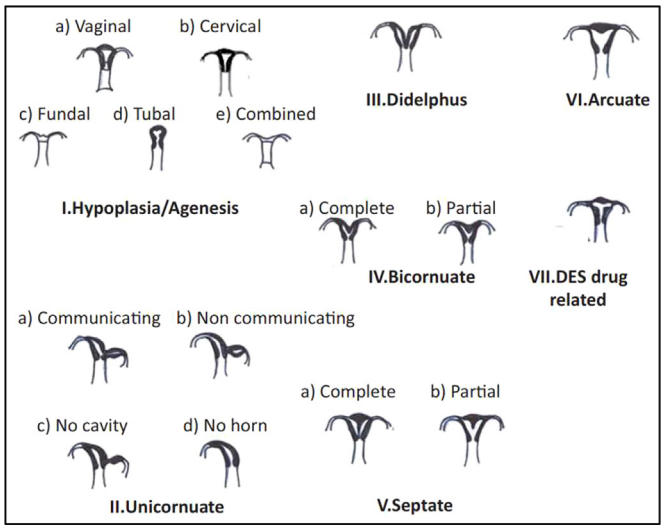

Fig. 3 – American Society for Reproductive Medicine classification [3].

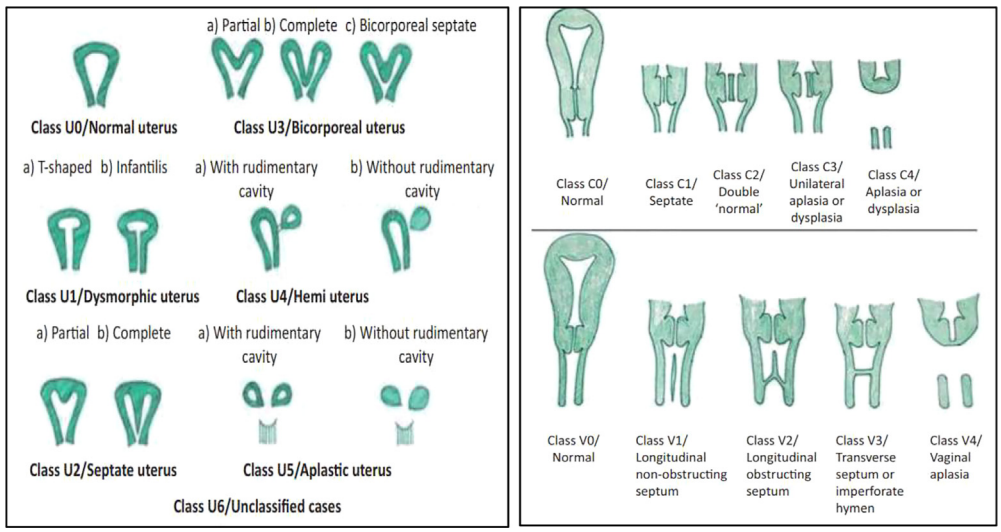

Fig. 4 – . The European Society of Human Reproduction and Embryology and European Society for Gynecological Endoscopy classification [3].

in (class I) of the ASRM and categorized as (U5a/C4/V4) in the ESHRE/ESGE classification [6]. Jegannathan et al. [3] classified a similar anomaly as (U6 – a mix of U3b, U4a, and U5a with C4 and V1/V3) according to the ESHRE/ESGE and (unclassified – Ie, IIb, and III mixed) according to the ASRM. Chandak et al. [7] state that our case could not be categorized into a specific category by the ASRM classification but could easily be categorized using the ESHRE/ESGE classification, which described as (U5a/C4/V4) [7].

Slightly different cases of 2 uterine horns, right rudimentary and left hematometra, with cervical atresia and blind short vaginal pouch, were reported by Dwivedi et al. [2] and classified as (U6/C4/V4) according to the ESHRE/ESGE without a proposed ASRM class. Others, like Bakri et al. [8] and Goluda et al. [9], described the anomaly as bicornuate, nonfused, separate rudimentary blind uterine horns with functioning endometria and complete cervicovaginal atresia or agenesis [8,9], where they could not assign it to any group of ASRM.

Some cases show vaginal or cervicovaginal atresia or agenesis with a normal uterine corpus or bulky uterus containing hemorrhagic endometrial collection and hematocervicometra with an absent vaginal opening [5,10].

Finally, published studies by Boruah et al. [11] and Sugi et al. [1] described similar cases under the spectrum of MRKH syndromes. They noted that most of these patients (92%) have unilateral or bilateral rudimentary uteri, with varying sizes and degrees of differentiation into 3 layers (ie, myometrium, junctional zone, and endometrium) [1]. As in our case, at T2WI, a low signal intensity fibrous band between the 2 horns was seen converging into the midline in these cases [1,11]. Considering this and our proposed classification for our case under (class Ie), according to ASRM, it might be under the broad spectrum of MRKH syndrome.

A comparison between these studies revealed that most authors easily and efficiently used the ESHRE/ESGE classification with similarity in the submitted class to their cases. Simultaneously, there was variability in the assigned ASRM classification, and some found it challenging to assign a specific class with this system. Worth mentioning is that some, like Jegannathan et al. and Chandak et al. [3,7], distinctly highlighted the superiority and simplicity of the ESHRE/ESGE classification, which is anatomical and embryological based and includes cervical and vaginal anomalies separately. Mahdavi et al. [6] added that more research projects on the pathophysiological and genetic aspects of these anomalies are required to unify MDA classification.

Another point of agreement among all researchers was that MRI had become a crucial and excellent diagnostic tool for those patients which accurately correlated with the surgical findings during exploratory laparotomies and laparoscopies as documented by Bakri et al., Sharma et al. and Boruah et al. [8,10,11].

Conclusion

MDA is widely variable and complex, with similar reported cases having identical findings described and classified differently by the authors. Thus, if the classification is uncertain using the known systems or a combined anomaly exists, it is better to give descriptive anatomical details of the abnormality rather than submitting single or multiple classes. Diagnostic imaging is essential for complete characterization and directs gynecologists' management plan, especially MRI.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

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