Imaging of Mayer-Rokitansky-Kü ster-Hauser (MRKH) Syndrome on MRI Examination in dr. Soetomo Hospital

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Abstract

Introduction: MRKH syndrome represents total or partial agenesis of the uterus and the two-thirds portion of the vagina, with 46XX karyotyping and normal bilateral ovaries, and a Fallopian tube. Despite the uterus agenesis, the Mullerian remnants, including uterine buds, fibrous bands, and midline triangular soft tissue, are visible. Magnetic Resonance Imaging (MRI) is a non-invasive modality with 100% accuracy, which is ideal for depicting Mullerian duct anomalies.

Materials and Methods: This is a descriptive study using a retrospective approach done in Dr. Soetomo General Hospital Surabaya and has obtained ethical permission. Our samples included 38 data from pelvic MRI acquired from medical records in Dr. Soetomo General Hospital between May 2018 and January 2022.

Results: The age group between 21 and 30 constituted a significant proportion of MRKH patients. The location of the right and left uterine buds relative to the ovarium were identified repeatedly in the anterior caudal, appearing isointense in the T2WI sequence. The right fibrous band was evident in 35 patients (94.7%), while the left fibrous band appeared in 35 patients (92.1%). Midline triangular soft tissue was visible in 35 patients (92.1%), showing a hypointense signal, 35 patients (92.1%) with intrapelvic right and left ovaries, and one patient (2.6%) with left endometrioma. The proximal vagina was not apparent in all patients, while 31 patients (81.6%) were shown to have the distal vagina.

Conclusion: MRI is can yield accurate information in describing anatomical abnormalities in MRKH. Although the findings of this study have been satisfactory, further research entailing larger samples is necessary to correlate the MRI features and surgery outcomes to assess its reliability.

Keywords: MRKH; MRI; uterine agenesis; Mullerian; fibrous bands

1. Introduction

The Müllerian duct is a significant component in forming the female internal reproductive organs. This process occurs in the early stages of embryo formation, requiring many intracellular and extracellular factors. Disruption of these form factors can result in multiple malformations and even death (Freire et al., 2020).

The Mullerian duct abnormalities entail a broad spectrum relying on the timeframe in which the disruption occurs during the development and differentiation process of the Mullerian duct. Mullerian duct development interruption is divided into three phases: formation, fusion, and absorption (Pitot, 2020).

The American Fertility Society (AFS) classified the Mullerian duct anomalies into seven classes. Class I refers to the Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome, making up roughly 15% of all Mullerian duct malformation (Bhayana and Ghasi, 2019). MRKH exhibits a set of clinical symptoms due to the failure of the paramesonephric (Mullerian) tract formation, happening in 1:4,500 baby girls (Wilson D and Bordoni B., 2021).

MRKH syndrome is characterized by total or partial agenesis of the uterus and the two-thirds portion of the vagina, in spite of 46XX karyotyping with normal bilateral ovaries and fallopian tubes. Even though the uterus fails to develop, the Mullerian residues are still evident, including the uterine buds (rudimental uterus), fibrous bands, and midline triangular soft tissue (Bhayana and Ghasi, 2019).

MRKH syndrome implies infertility and sexual intercourse disruption during reproductive age (Hall-Craggs et al., 2013). A study conducted by Chen N et al. (2021) found that the mean age of 1,055 MRKH patients during their prior visits to 11 health centers was 24.8 (21.3 – 29.0) years old. The most frequent symptoms presented by the patients were primary amenorrhea in 969 subjects (91.8%); 71 patients (6.7%) came with dyspareunia; the remaining 15 (1.4%) complained of primary amenorrhea with chronic and cyclic abdominal pain (Chen N et al., 2021).

MRKH patients with uterine agenesis have around 40% of the endometrium. Approximately 50% have experienced menstrual cycle symptoms, such as pelvic pain, although without bleeding. Endometriosis may occur in patients with MRKH; therefore, therapeutic managements are requisite, one of which is surgery (Tian W et al., 2021).

MRI is an imaging modality in a non-invasive manner with advanced accuracy and hence is excellent in diagnosing Mullerian duct malformation (Narang, Cope, and Teixeira, 2018). In addition, MRI may visualize the involvement of the neighboring organs, for instance, ovaries, kidneys, urinary tracts, and bone deformation (Maciel et al., 2020).

2. Methods

The authors investigated the patients in Dr. Soetomo General Hospital from May 2017 until January 2022. The medical records that met the inclusion criteria were considered for the research subjects, and their data were obtained, including the MRI results, which subsequently were blindly evaluated by a breast and female imaging consultant.

Subjects were taken using consecutive sampling where every patient who met the inclusion criteria was included in this study during the specified period.

2.1 Inclusion and Exclusion Criteria

2.1.1 Inclusion Criteria

- MRKH patients who underwent MRI examination at RSUD Dr. Soetomo before taking action
- Patients confirmed MRKH either from physical examination or post-op reports and have had an MRI examination

2.1.2 Exclusion Criteria

- Patient data with primary amenorrhea with other causes
- Patients who have had neovaginal surgery

The observed variables in this study comprised segmental MRI images distributed according to the American Society of Reproductive Medicine (ASRM) classification for MRKH patients (Bhayana and Ghasi, 2019):

2.2 The presence of the uterine buds

- If present, is it unilateral or bilateral, and how is the signal intensity in T2WI
- Location
- The existence of cavitations and intraluminal hemorrhage
- The presence of a fibrous band connecting the uterine buds

2.3 Vagina

- The presence of the proximal and distal vaginal which are separated embryologically. The vagina is a hyperintense mucosa circled by hypointense fibromuscular walls with hyperintense paravaginal vein plexus.

2.4 Ovaries

- The presence of the ovaries
- Location, whether it is intra- or extra-pelvic. The ovaries are said to be extra-pelvic should they are situated in the abdomen, inguinal canals, and anterior or anterolateral sites of iliac arteries and psoas muscles.

2.5 Coexisting abnormalities that may be evident, such as kidney and vertebral anomalies.

3. Results

This research was an observational study aiming to illustrate the classification of MRKH segmentation based on the MRI examination. The qualified samples in this study included 38 subjects, with ages ranging from 14 to 43, with an average of 24.

Table. 1 Basic characteristics of the subject

Age group (years)	Frequency	Percentage (%)
10 – 20	13	34,1
21 – 30	20	52,6
31 – 40	4	10,6
41 – 50	1	2,7
Total	38	100.0

Table. 2 The result of the MRKH segmentation on pelvic MRI examination

1.	Right and left uterine buds craniocaudal position within related to ovary	Frequency	Percentage (%)
	absent	6	15.8
	cranial	1	2.6
	same	6	15.8
	caudal	25	65.8
	Total	38	100.0
2.	Right and left uterine buds anteroposterior position within related to ovary		
	absent	6	15.8
	anterior	19	50.0
	same	9	23.7
	posterior	4	10.5
	Total	38	100.0

3. Right and left uterine buds signal intensity		
absent	6	15.8
Hypointense	3	7.9
Isointense	28	73.7
Hyperintense	1	2.6
Total	38	100.0
4. Right and left fibrous band signal intensity		
absent	2	5.3
Hypointense	35	92,1
Isointense	1	2,6
Hyperintense	-	-
Total	38	100.0
5. Midline triangular soft tissue signal intensity		
absent	3	7.9
Hypointense	-	-
Isointense	35	92,1
Hyperintense	-	-
Total	38	100.0
6. Right and left ovary		
absent	3	7.9
present	35	92.1
Total	38	100.0
7. Right and left ovary location		
absent	3	7.9
Intrapelvis	35	92.1
Extrapelvis	-	-
Total	38	100.0
8. Right ovarian mass		
absent	38	100.0
present	-	-
Total	38	100.0
9. Left ovarian mass		
absent	37	97.4
present	1	2.6
Total	38	100.0
10. Proximal vagina		
absent	38	100
present	-	-
Total	38	100.0
11. Distal vagina		
absent	7	18.4
present	31	81.6
Total	38	100.0

4. Discussion

In this study, our sample distribution conforming to the age group was comprised of 13 patients (34.1%) aged between 10 to 20, 20 patients (52.6%) between 21 to 30, four patients (10.6%) between 31 to 40, and one patient (2,7%) between 41 to 50 (table.1).

4.1 Uterine buds

There were 32 patients (84.2%) with both right and left uterine buds, while the remaining 6 (15.8%) did not show the presence of the uterine buds. The right and left uterine buds were recurringly situated caudally with respect to the ovarium. Most anteroposterior right and left uterine buds relative to the ovarium were located at the anterior site (Fig. 1), with isointense signals at the T2WI sequence frequently occurring among these subjects.

This discovery aligned with a prior study by Bhayana (2019), who stipulated that the right and left uterine buds, according to the craniocaudal position to the ovarium, are mainly situated at the caudal site; in consonance with the anterior position, they occur most often at the anterior site.

This assumption was in accordance with a theory stating that the Mullerian duct will migrate, elongate, and form the Mullerian canal sliding lateral to the mesonephric duct, crossing ventrally and developing caudomedially. In the middle portion, the duct will proximate with the paramesonephric from the opposite (Laterza, 2021).

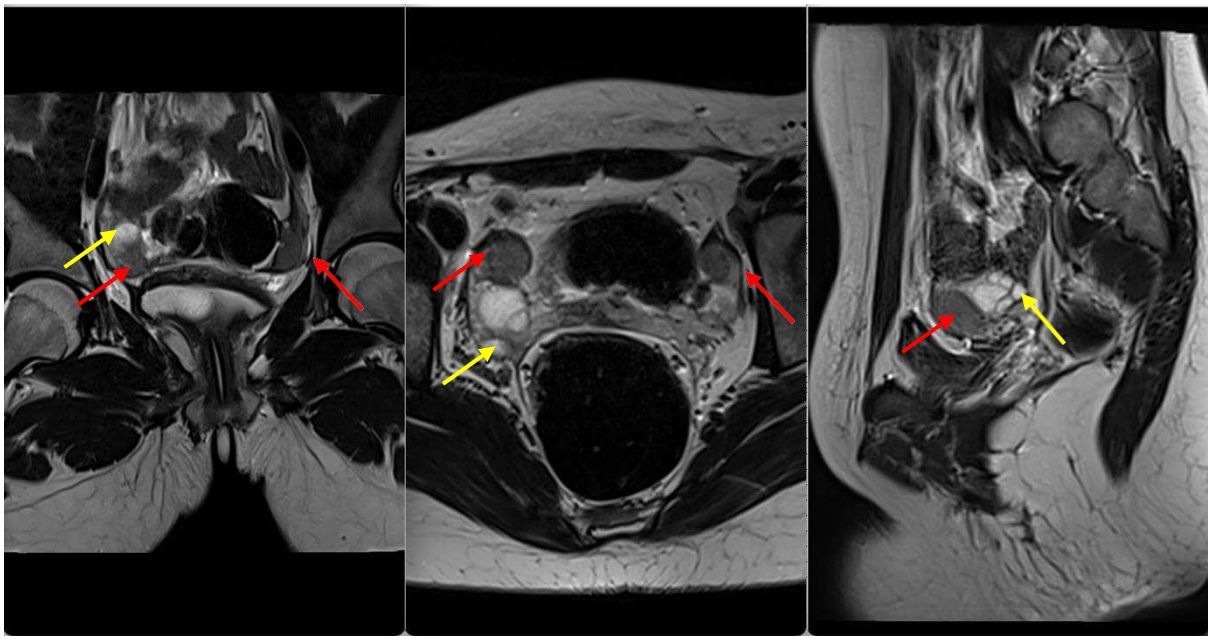

Fig. 1. The right and left uterine buds (red arrows) are placed at the caudal and anterior position to the ovarium (yellow arrows) with a hypointense signal at the T2WI sequence (Courtesy of Dr. Soetomo Hospital).

4.2 Fibrous bands

Fibrous bands represent a structure connecting the bilateral uterine buds. The right fibrous band existed in 36 subjects (94.7%) and did not develop in two patients (5.3%), appearing hypointense entirely in the T2WI sequence (Fig. 2).

The left fibrous band was evident in 35 patients (92.1%), while the remaining 3 (7.9%) did not have the left fibrous band, with most signal intensity displayed as hypointense in 33 patients (86.8%) at the T2WI sequence (Figure 2). This was in line with a study conducted by Yoo Roh-Eul et al., 2013, who specified that the majority of right and left fibrous bands appeared hypointense at the T2WI sequence.

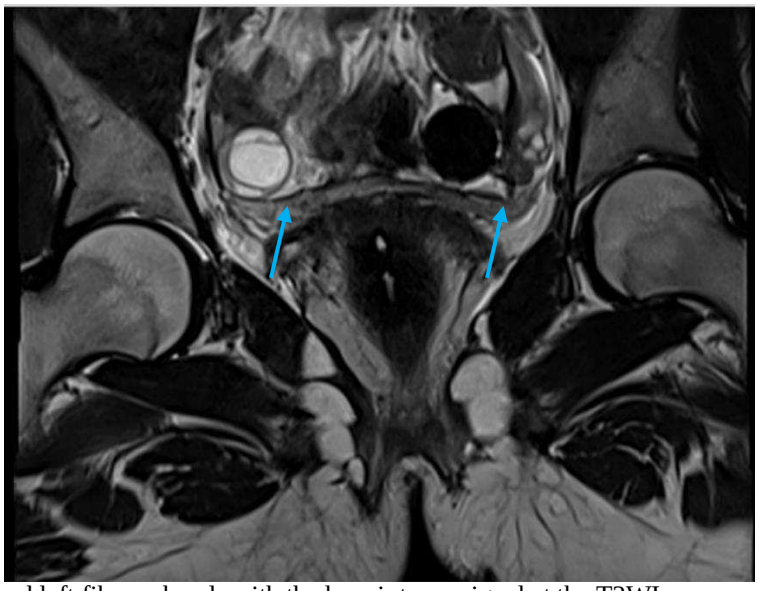

Fig. 2. The right and left fibrous bands with the hypointense signal at the T2WI sequence (Courtesy of Dr. Soetomo Hospital)

4.3 Midline Triangular Soft Tissue

The midline triangular soft tissue is a structure formed by both rights and left fibrous bands stationed at the center in the triangular shape. There were 35 subjects (92.1%) having midline triangular soft tissue, while three samples (7.9%) did not display the structure. The signal intensity of all midline soft tissues was hypointense at the T2WI sequence (Fig. 3a). This result was discordant with a previous study by Bhayana (2019), in which 13 subjects out of 18 appeared hyperintense and five patients hypointense at the T2WI sequence.

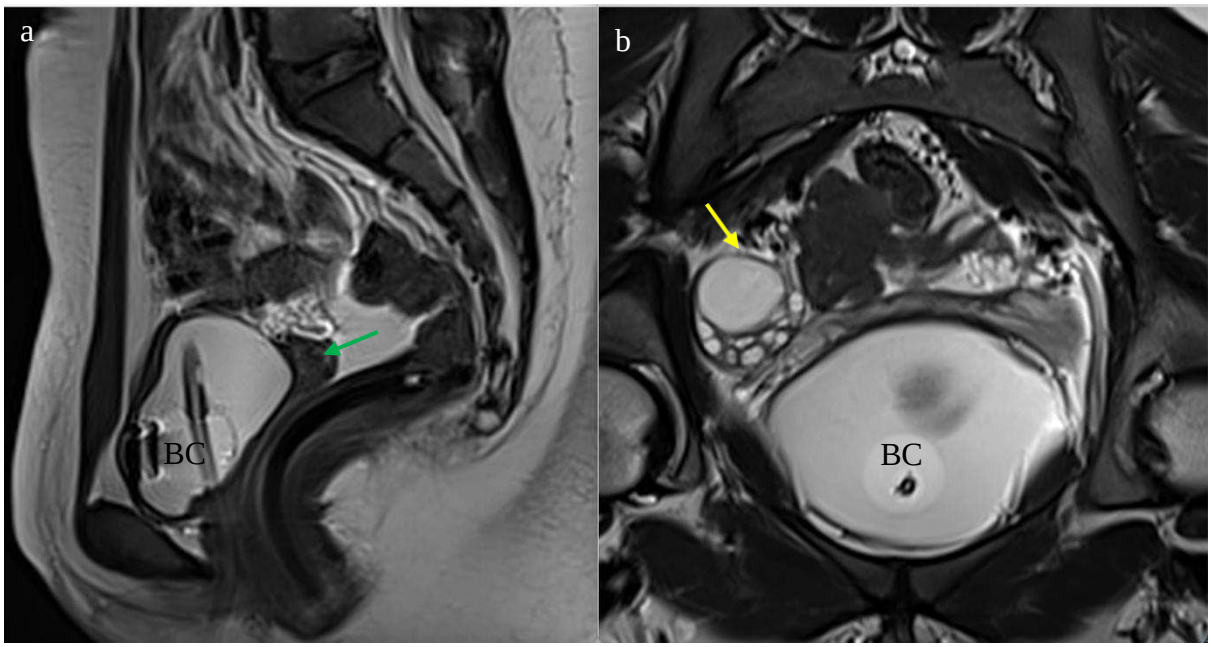

Fig. 3. (a) Midline triangular soft tissue (green arrow); (b) Ovarium (yellow arrow); BC (Balloon Catheter) (Courtesy of Dr. Soetomo Hospital)

4.4 Ovarium

Our study result showed that both right and left ovaries were evident in 35 patients (92.1%), all positioned within intrapelvic (Fig. 3b), whereas it did not appear in three patients (7.9%). A similar finding was outlined by Yoo Roh-Eul et al. (2013), stating that all ovaries were situated in the pelvic cavity.

One patient (2.6%) appeared to have endometriosis in her left ovary. Endometriosis is defined by an endometrium tissue developing outside the uterine cavity, eliciting a chronic inflammatory reaction and disturbing the reproductive process. There is a myriad of papers detailing the origin of endometriosis (Fig. 4).

Coelomic metaplasia (coelomic epithelial cells covering the ovaries and peritoneum transform into endometrial cells), or ectopic primitive cell growth outside the Müllerian ducts, are the suspected source of endometriosis. Other theories, such as the transformation of circulating stem cells into endometrial tissue, are also considered (Bourgioti C et al., 2017).

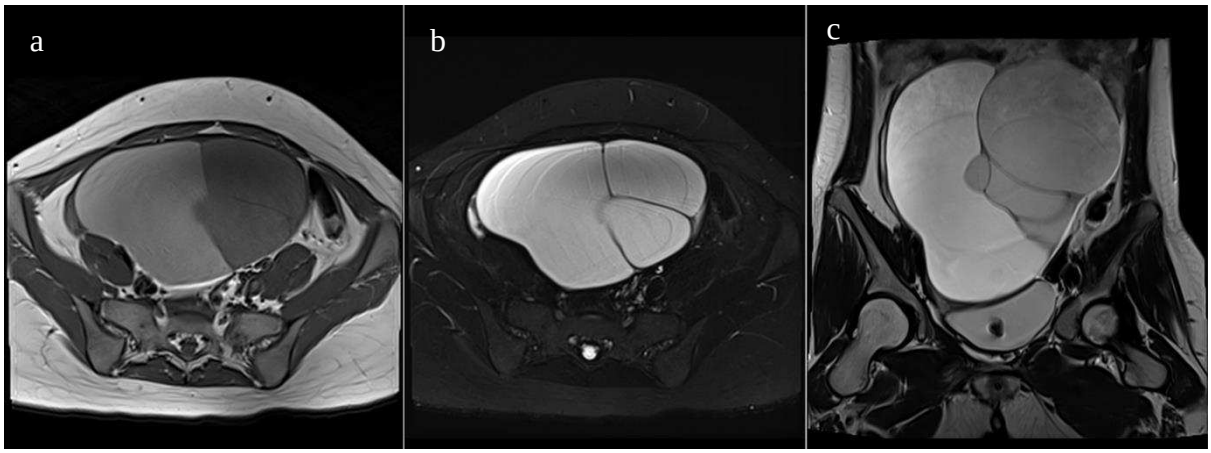

Fig. 4. A 19-year-old female with primary amenorrhea and abdominal pain. Pelvic MRI shows the left endometrioma and hemosalpinx (a), which appears hyperintense at the T1WI; (b) hyperintense at T2WI Fat Sat, T2 Shading (+); (c) coronal T2WI (Courtesy of Dr. Soetomo Hospital)

4.5 Vagina

All subjects did not have the proximal vagina. There were 31 samples (81.6%) showed the distal vagina (Fig. 5); meanwhile, it did not exist in the remaining 7 (18.4%). This result was supported by Deb Kumar Boruah et al. (2013) and Bhayana (2019), stating the absence of a proximal vagina.

The Mullerian ducts began to be absorbed and gradually developed into the uterine cavity, cervix, and upper vagina following week 12 to week 14, and then the sinovaginal originating from the urogenital sinus will become the distal vagina (Pitot, 2020). The insufficiency of Mullerian duct evolution will instigate agenesis or hypoplasia of the two-thirds of the proximal vagina, cervix, and uterus (Bhayana and Ghasi, 2019).

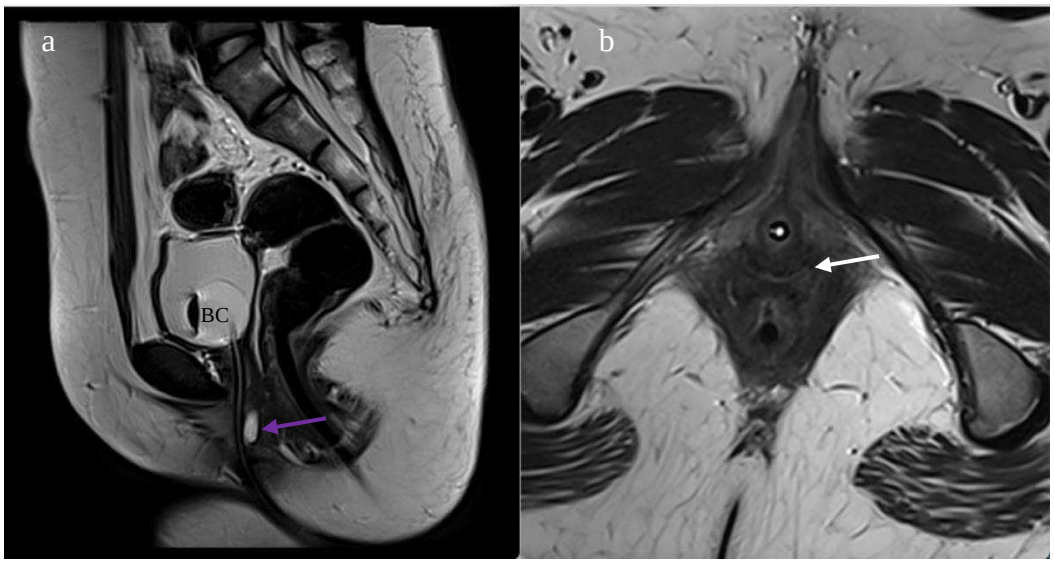

Fig. 5 (a,b). Distal vagina (white arrow), without the proximal vagina. BC (Balloon Catheter); M (marker at the vaginal introitus, purple arrow) (Courtesy of Dr. Soetomo General Hospital)

5. Conclusion

This study did not compare with surgery in order to compare anatomical structures based on segmentation classification in MRKH patients. Although the findings of this study have been satisfactory, further research entailing larger samples is necessary to correlate the MRI features and surgery outcomes to assess its reliability.

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