Contents lists available at IOCS

Science Midwifery

journal homepage: www.midwifery.iocspublisher.org

Hematometra ec agenesis serviks, suspek MRKH

Selvy Anriani[1], Prastika Candra Triastuti[2], Gde Ngurah Bayu Dalem Putra[3]

[1]General Practitioners, Nusa Tenggara Timur, Indonesia

[2]General Practitioners, Special Region of Yogyakarta, Indonesia

[3]GeneralPractitioners, Bali, Indonesia

ARTICLE INFO

Article history:

Received Dec 18, 2023
Revised Dec 19, 2023
Accepted Dec 30, 2023

Keywords:

Cervical Agenesis
Hematometra
MRKH

ABSTRACT

Background: Primary amenorrhea is the absence of menarche. After gon-adal dysgenesis, the Mayer-Rokitansky-Küster-Hauser (MRKH) syn-drome is the second most common cause of primary amenorrhea. Herein, we de-scribe how to diagnose and the treatment of MRKH. Case presentation: A 21-year-old lady with complaints of abdominal pain in the lower portion of the navel for the past 4 months that has worsened in the previous week, accompanied by complaints of urinat-ing slowly and feeling agony. The patient claimed to be single, to have never had coitus, and to have never menstruated previously. Physical examination revealed tanned 5 breasts, axillary hair (+), and an ab-dominal lump palpable on the 2nd finger region below the navel with pain. There were 5 tanner pubic hairs, labia major +/+, labia minor +/+, and a hymen bulging (-) in the vagi-nal area. A cystic mass pressing on the anterior region of the rectum, hy-men (+) impression of the vaginal septum, and vaginal introitus (-) are all felt when examined in the anus. Uterine dimensions 10x5x4 cm, hypoechoic mass in the uterine cavity, free fluid (-), cervical canal not evident, vaginal introitus (-), hematocol-pos (-), and abdominal ultrasonography results re-vealed no abnormali-ties. Other tests were not carried out. A laparotomy was conducted to remove the hematometra based on the results of clinical and support-ive investigations. Conclusion: This is a rare occurrence of cervical agenesis, occurring in 1 in 80,000 women. Additional tests are required in this case to con-firm the diagnosis of MRKH. The treatment in this scenario seeks to reduce pain and evacuate blood, so that subsequent treatment to re-store the cervical canal at a more comprehensive health service cen-ter is required.

Corresponding Author:

Selvy Anriani,
General Practitioners,
Karitas Hospital,
Jl. Sapurata, Wee Tobula, District. Loura, Southwest Sumba Regency, East Nusa Tenggara, Indonesia
Email: selvy58.sa@gmail.com

INTRODUCTION

The Mayer Rokitansky Kuster Hauser (MRKH) syndrome is a female reproductive system disorder defined by the loss or disappearance of the uterus and vagina arising from the Mullerian ducts, but

the ovaries function correctly (Mayuri et al., 2014). The causes of the aforementioned conditions are unknown. The incidence is extremely low, at one in every 4500 to 5000 women (Yeni, 2020). MRKH with vaginal agenesis is the most prevalent form, while cervical agenesis is a rare incidence with a 1:80,000 - 100,000 occurrence rate (Dwivedi et al., 2020; Pratiwi & Fauzi, 2018).

Primary amenorrhea with normal thelarche and adrenarche, as well as abnormalities in sexual intercourse and infertility, are characteristic clinical signs of MRKH syndrome (Muñoz et al., 2011). MRKH is categorized into three types: type I is the presence of anomalies in the reproductive organs (utero-vaginal aplasia), type II is the presence of asymmetric utero-vaginal aplasia or hypoplasia followed by hypoplasia or the absence of one or both fallopian tubes, and type III is the absence of one or both fallopian tubes, type III or MURC (Mullerian duct aplasia, Renal dysplasia, and Cervical somite abnormality) are urological anomalies that occur in 15-40% of cases, skeletal anomalies in 20-40% of cases, and hearing, cardiac, and digitalis problems are infrequent (Kraiem et al., 2020).

Physical, laboratory, and supportive exams such as ultrasonography, MRI, laparoscopy, and pyelography are commonly used to make a diagnosis (Miyake et al., 2023). When secondary sexual growth was discovered during normal puberty and normal height, a speculum examination may be performed to see the vaginal area, however this was problematic in certain cases because most MRKH disorders were identical to vaginal agenesis. Typically, the vulva, labia majora, minora, and clitoris are found to be within normal limits (Dwivedi et al., 2020). Chromosome analysis to rule out X chromosome karyotype abnormalities and AIS (androgen insensitivity syndrome) are two examples of laboratory procedures. Ultrasonography (ultrasound), MRI, laparoscopy, and pyelography are commonly used in diagnosis. In this scenario, MRI is the gold standard for assessment since it can be utilized to validate inconclusive ultrasound results linked to uterine cavitation (Pluta et al., 2020).

Treatment of MRKH syndrome with vaginal drugs can be done non-surgically or surgically. The Frank approach is used for non-surgical care, whereas surgery can be performed using the McIndo technique, Williams Vaginoplasty, rotating flap procedure, bowel neovaginal technique, and Vecchieti technique (Ng et al., 2020; Thomas et al., 2015). In most cases of MRKH with cervical agenesis, minimally invasive therapy, such as laparoscopy with continued fertility in mind, or a laparotomy to remove the hematometra, can be used (N Fobellah et al., 2021; Zhang et al., 2017).

This study contributes to knowledge and management skills for diagnosis and treatment of MRKH cases based on real cases that occur in the field.

RESEARCH METHOD

Research design with Case Report (CARE) which describes patient case problems that are considered to have scientific value. There are cases associated with Hematometra ec agenesis of the cervix, suspected cases of MRKH.

A 21-year-old lady with complaints of abdominal pain in the lower portion of the navel for the past 4 months that has worsened in the last week, accompanied by complaints of urinating slowly and feeling agony. The patient claimed to be single, to have never had coitus, and to have never menstruated previously. Physical examination revealed tanned 5 breasts, axillary hair (+), and an abdominal lump palpable on the 2nd finger region below the navel with pain. There were 5 tanner pubic hairs, labia major +/+, labia minor +/+, and hymen bulging (-) in the genitalia area. There is a cystic mass pushing on the anterior section of the rectum, hymen (+) impression of the vaginal septum, and vaginal introitus (-) when inspected in the anus.

Routine blood laboratory, urinalysis, and abdominal and obstetric ultrasound were performed as support examinations. Routine blood tests revealed no abnormalities, but urine revealed germs and full leukocytes. Ultrasound test results revealed a hypoecho-ic mass in the uterine cavity, free fluid (-), a cervical canal that was not apparent, vaginal introitus (-),

hematocolpos (-), and no abnormalities in the abdominal ultrasound. Other supporting tests were not performed.

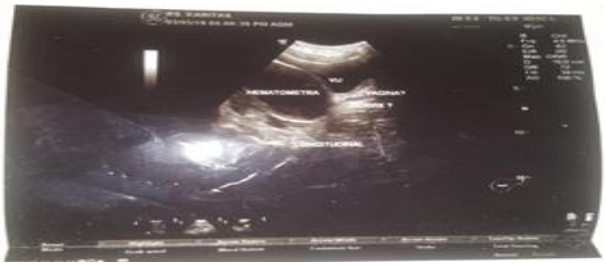

Figure 1. Obstetric ultrasound results

A laparotomy was conducted to remove the hematometra based on the findings of the clinical and supportive exams. During the laparotomy, 300 cc of blood clots were emptied, an anteflexed uterus was observed, and no internal uterine ostium was discov-ered.

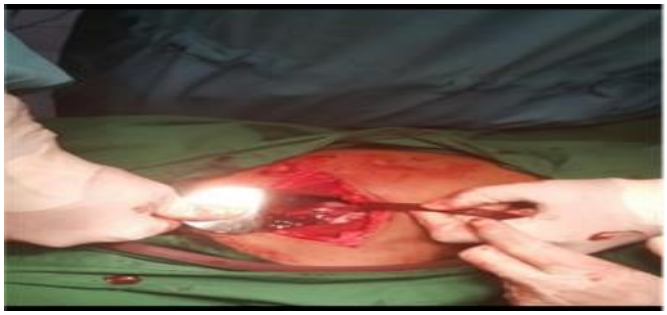

Figure 2. Hematometra expulsion via laparotomy

RESULTS AND DISCUSSIONS

In general, MRKH syndrome is divided into two types, with type I characterized by isolated cases of utero-vaginal agenesis and type II characterized by cases of utero-vaginal agenesis associated with extra-genital anomalies, such as urologic (e.g. renal agenesis, pelvic kidney, and horseshoe kidney), skeletal, auditory, and cardiac anomalies (Filho et al., 2019). The most severe form of type II MRKH syndrome is the so-called Müllerian hypoplasia, renal agenesis, cervicothoracic somite dysplasia (MURCS) association (Ledig & Wieacker, 2018).

According to the American Fertility Society, the aforesaid situation is rare and difficult to cure, especially suspicion of mrkh syndrome with cervical agenesis (Joshi et al., 2022; Zaidi et al., 2017). In this patient, with complaints of not having menstruation until the age of 21 years, accompanied by complaints of severe abdominal pain without other complications or pain, and secondary sexual growth within normal limits, MRKH type one syndrome can be suspected, which is only followed by agenesis of the reproductive organs without complaints or other complications (Hafid et al., 2023; N Fobellah et al., 2021). In cases of MRKH syndrome, the diagnosis must be supported by other tests, such as chromosomal analysis or AIS, and an MRI may be recommended to enhance the findings from the ultrasound results. However, due to inadequate facilities in rural places, it was not performed on this patient (Fairuza, 2022). The care of patients with the aforementioned disorders is still controversial and has numerous variations due to several factors related to the patient's desire to continue having children (Rezai, 2017; Thorne et al., 2018). Some individuals are recanalized via the endocervical canal to allow menstrual blood to flow while still protecting the uterus, or cervical-uterine recanalization can also be accomplished using laparotomy

or laparoscopy (Afodun et al., 2020). Minimally invasive therapy, such as laparoscopy, is also mentioned as a good therapeutic choice with a better prognosis (Pizzo et al., 2013).

Ultrasound examination is frequently the initial diagnostic test in the evaluation of patients with MRKH syndrome; it can confirm the presence of ovaries and lack of uterus (Londra et al., 2015). Nevertheless, due to technical issues, the results are occasionally inconclusive. MRI is the preferred imaging modality for confirming the diagnosis and identifying any associated malformations (Al Dandan et al., 2019). Preibsch et al. have demonstrated an excellent relationship between MRI and laparoscopy findings in patients with MRKH syndrome (Herlin et al., 2020).

In this patient's situation, a laparotomy surgery was conducted to remove blood clots or evacuate the hematometra, as well as to lessen and relieve the patient's suffering. The patient will then be referred to a larger hospital for a more thorough assessment and care for further treatment. The patient was referred to Sanglah General Hospital one month later.

CONCLUSION

This study is a basic guide in efforts to develop standard operational procedures for MRKH diagnosis and treatment management. This study used primary data were involved 1 MRKH patient who has received diagnostic management and treatment according to procedures. Next, the researchers carried out a retrospective data analysis by looking at the patient's current health condition which was related to previous interventions. This is an extremely unusual instance with cervical agenesis suspected of being MRKH syndrome, thus proper inves-tigation and treatment are required. Treatment in this scenario is still highly debated, but various factors must be examined in order to give proper care, including the patient's wish to continue having children. Based on these consid-erations, a low-risk therapy with a long-term evaluation can be chosen.

The diagnosis of MRKH syndrome imposes a significant psychological burden on patients due to the associated infertility. Through psychological counseling and support groups, the distress can be alleviated. The condi-tion can be treated with progressive vaginal dilators or surgical creation of a neovagina. Fertility options may include assisted reproductive technology and surrogacy.

ACKNOWLEDGEMENTS

The researchers acknowledge the Research and Innovation Institute of Karitas Waitabula Hospital, which has funded the entire process of this research activity.

References

Afodun, A. M., Ukwenya, V. O., Quadri, K. K., Malinga, R. J., Ahimbisibwe, O., & Wandabwa, J. (2020). Sonographic presentation acute hematometra and massive hematocolpos in a pubertal girl: A case report. *Journal of Biomedical Sciences*, *7*(2), 58–63. https://doi.org/10.3126/jbs.v7i2.33999

Al Dandan, O., Hassan, A., Alsaihati, A., Aljawad, L., & Almejhim, F. (2019). A rare form of Mayer-Rokitansky-Küster-Hauser syndrome: Case report and review of literature. *Case Reports in Women's Health*, *24*, e00137. https://doi.org/10.1016/j.crwh.2019.e00137

Dwivedi, D., Jain, M., Jain, S., & Jain, S. (2020). A rare case report on complete cervical agenesis with vaginal atresia and suspended didelphys uterus with hematometra and left haematosalpinx. *International Journal of Reproduction, Contraception, Obstetrics and Gynecology*, *9*(3), 1274. https://doi.org/10.18203/2320-1770.ijrcog20200914

Fairuza, F. (2022). Haematocolpos and Secondary Hematometra Due to Imperforate Hymen: Diagnosis and Treatment. *Jurnal Biomedika Dan Kesehatan*, *5*(3), 228–235. https://doi.org/10.18051/JBiomedKes.2022.v5.228-235

Filho, L. F. D. P., Neves, G. C., Bandoli, L. F., Coelho, L. C. D. A., Feital, N. D. S. F., Murad, R. V., Jesus, A. L. C. D., Sá, D. A. D., Sá, L. B. C. D., Mwambire, J., & Arbex, A. K. (2019). Case Report: Clinical and

Diagnostic Aspects of Mayer-Rokitansky-Kuster-Hauser Syndrome. *Health*, *11*(10), 1367–1372. https://doi.org/10.4236/health.2019.1110105

Hafid, A., Rodiani, R., & Sayuti, M. (2023). Amenorea Primer et Hematokolpos et Hematometra ec. Septum Vagina Transversal. *Jurnal Ilmiah Kesehatan Sandi Husada*, *12*(1), 55–60. https://doi.org/10.35816/jiskh.v12i1.867

Herlin, M. K., Petersen, M. B., & Brännström, M. (2020). Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome: A comprehensive update. *Orphanet Journal of Rare Diseases*, *15*(1), 214. https://doi.org/10.1186/s13023-020-01491-9

Joshi, S., Bhandari, A. R., Shrestha, P., & Shakya, R. (2022). Dissociative disorder in Mayer Rokitansky Küster Hauser syndrome with pulmonary agenesis: A case report. *The Egyptian Journal of Neurology, Psychiatry and Neurosurgery*, *58*(1), 3. https://doi.org/10.1186/s41983-021-00440-y

Kraiem, S., Zoukar, O., Hnayin, A., Zouari, A., Faleh, R., & Haddad, A. (2020). Complete cervical agenesis: Successful surgical treatment: One case report. *Pan African Medical Journal*, *36*(211), 1–4. https://doi.org/10.11604/pamj.2020.36.211.24408

Ledig, S., & Wieacker, P. (2018). Klinische und genetische Aspekte des Mayer-Rokitansky-Küster-Hauser Syndroms. *Medizinische Genetik*, *30*(1), 3–11. https://doi.org/10.1007/s11825-018-0173-7

Londra, L., Chuong, F. S., & Kolp, L. (2015). Mayer-Rokitansky-Kuster-Hauser syndrome: A review. *International Journal of Women's Health*, *7*, 865–870. https://doi.org/10.2147/IJWH.S75637

Mayuri, J., Vasudha, N., Vijay, G., Asha, P., & J, P. (2014). Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome: A Case Report. *Journal of Research in Medical and Dental Science*, *2*(1), 106. https://doi.org/10.5455/jrmds.20142120

Miyake, A., Kobayashi, Y., Imaeda, K., Yoshihama, T., Nakamura, K., Yokota, M., Hayashi, S., Yamagami, W., Banno, K., & Aoki, D. (2023). Case Series of Mayer–Rokitansky–Küster–Hauser Syndrome: Analysis of 17 Cases. *Clinical and Experimental Obstetrics & Gynecology*, *50*(1), 1. https://doi.org/10.31083/j.ceog5001001

Muñoz, M. del M., Noguero, R., & Martín, S. (2011). Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome. Diagnostic and therapeutic approach of a rare disease. *Colombia Medica*, 369–372. https://doi.org/10.25100/cm.v42i3.884

N Fobellah, N., Asahngwa, C., Bongfen, C., Gobina, R., Odette, K., Ngwa, W., & Foretia, D. (2021). Hematometra in a Patient without Previous Surgeries/Anomalies in a Resource Constraint Setting-A Case Report from Bangem District Hospital. *Archives of Clinical and Biomedical Research*, *05*(03). https://doi.org/10.26502/acbr.50170173

Ng, K., Ip, P. N., Yiu, K., Chung, J. P., & Chan, S. S. (2020). Treatment of patients with Mayer-Rokitansky-Küster-Hauser syndrome in a tertiary hospital. *Hong Kong Medical Journal*. https://doi.org/10.12809/hkmj208467

Pizzo, A., Laganà, A. S., Sturlese, E., Retto, G., Retto, A., De Dominici, R., & Puzzolo, D. (2013). Mayer-Rokitansky-Kuster-Hauser Syndrome: Embryology, Genetics and Clinical and Surgical Treatment. *ISRN Obstetrics and Gynecology*, *2013*, 1–10. https://doi.org/10.1155/2013/628717

Pluta, D., Lemm, M., Franik, G., Kowalczyk, K., Blukacz, Ł., Tekieli-Balon, A., & Madej, P. (2020). Mayer-Rokitansky-Küster-Hauser syndrome – case studies, methods of treatment and the future prospects of human uterus transplantation. *European Review for Medical and Pharmacological Sciences*, *24*, 549–563.

Pratiwi, R., & Fauzi, A. (2018). Laporan kasus: Neovagina Ileum Pada Agenesis Vagina Wanita Dewasa. *Syifa' MEDIKA*, *8*(2), 90–94.

Rezai, S. (2017). Three Cases of Didelphys Uterus, Including Uterus Didelphys with Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA Syndrome) AKA Herlyn-Werner-Wunderlich Syndrome), with a Systematic Review. *Obstetrics & Gynecology International Journal*, *7*(2). https://doi.org/10.15406/ogij.2017.07.00241

Thomas, E., Shetty, S., Kapoor, N., & Paul, T. V. (2015). Mayer-Rokitansky-Kuster-Hauser syndrome. *BMJ Case Reports*, 1–2. https://doi.org/10.1136/bcr-2015-210187

Thorne, J. G., Russell, E. H., Rumbolt, D., & Jamieson, M. A. (2018). Cause or Coincidence? Spontaneous Hematometra in Young Women Receiving Depomedroxyprogesterone Acetate: A Small Case Series. *Journal of Pediatric and Adolescent Gynecology*, *31*(4), 416–419. https://doi.org/10.1016/j.jpag.2018.01.005

Yeni, D. (2020). *Mayer-Rokitansky-Küster-Hauser syndrome Cut Meurah Yeni , 2 Khairussani , 3 Wardatul Bararah*. *20*(1), 47–52.

Zaidi, M. S., Hassan, A., & Almogbel, E. (2017). Mayer-Rokitansky-Küster-Hauser Syndrome In A Young Woman. *AACE Clinical Case Reports*, *3*(2), e93–e95. https://doi.org/10.4158/EP151105.CR

Zhang, Y., Chen, Y., & Hua, K. (2017). Outcomes in patients undergoing robotic reconstructive uterovaginal anastomosis of congenital cervical and vaginal atresia. *International Journal of Medical Robotics and Computer Assisted Surgery*, *13*(3), 1–6. https://doi.org/10.1002/rcs.1821