A Rare Case of Mayer-Rokitansky-Kuster-Hauser (MRKH) Syndrome with Inguinal Herniation of the Ovary: Case Report and Review of Literature

Medha Davile*, Amrita Singh, Lanka Vasudha Vani and Shuchita Mundle

Department of Obstetrics and Gynecology, All India Institute of Medical Sciences, Nagpur, India

Received: 26 September 2024

Accepted: 8 December 2024

*Corresponding author: nagpure.medha@gmail.com

DOI 10.5001/omj.2028.03

Abstract

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is a rare congenital disorder characterized by the absence or underdevelopment of the uterus and upper vagina, with normal ovarian function. Inguinal herniation of the ovary is an uncommon clinical finding, particularly in adult females, and is often associated with pediatric patients. The combination of ovarian herniation and MRKH syndrome is even rarer and likely under-reported. We present the case of a 19-year-old female with MRKH syndrome Type II, who presented with a right inguinal hernia. Imaging revealed the herniation of the right ovary, a rudimentary fallopian tube, and a rudimentary uterus. Surgical exploration confirmed these findings, and the hernia was repaired with mesh placement. The patient's postoperative recovery was uneventful, and she was counselled regarding future reconstructive options and fertility prospects. This case highlights the importance of considering underlying congenital anomalies such as MRKH syndrome in cases of ovarian herniation. Early diagnosis and surgical intervention are crucial to prevent complications such as ovarian torsion and infarction. A review of the literature underscores the rarity of this condition and the need for awareness among clinicians to ensure timely management.

Keywords: Mayer Rokitansky Kuster Hauser Syndrome; MRKH; Inguinal Hernia; Herniated Ovary; Congenital Anomaly; Herniorrhaphy; Vaginoplasty.

Introduction

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a rare congenital disorder in karyotypic females (46, XX) characterized by the underdevelopment or absence of the uterus and vagina. Despite these anomalies, the ovaries, external genitalia, and secondary sexual characteristics typically develop normally.[1] Girls with MRKH syndrome often present with primary amenorrhea. It is the second most common cause of primary amenorrhea after hypogonadism.[2]

MRKH syndrome is categorized into two subtypes[3,4]:

- **Type I**, involving malformations limited to the genital system.
- **Type II**, associated with additional abnormalities, such as renal and skeletal anomalies.

Herlin et al.[5] conducted a nationwide registry-based study to describe the prevalence and characteristics of MRKH syndrome. Common genitourinary complaints included amenorrhea, lower abdominal pain, and dyspareunia, with kidney malformations being the most frequently observed extragenital anomalies. Herniation of the rudimentary uterus and ovary is a rare association with MRKH syndrome, with only few cases reported in the literature.

Inguinal hernia (IH) is one of the most common types of hernia in adult males but is less frequently observed in females.[6] In most cases, the inguinal hernia sac contains omentum, small intestine, appendix, or even the urinary bladder.[6] However, herniation of the ovary in adult females is a rare occurrence, more commonly seen in infants and pediatric patients.[6] The presence of the uterus within an inguinal hernia is particularly uncommon.[7] When encountering ovarian, fallopian tube, or uterine herniation, it is crucial to investigate for underlying congenital anomalies, such as Mullerian duct anomalies and Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome.[6,7]

The following case report details the presentation, diagnosis, and surgical management of a young woman with MRKH syndrome who presented with an inguinal hernia containing ovary.

Case Report

A 19-year-old unmarried female presented to the outpatient department with a swelling in her right inguinal region, persisting for 3 to 4 months, accompanied by recent onset of pain. She had a history of primary amenorrhea and had been previously diagnosed with Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome. The patient exhibited a female phenotype, with well-developed axillary and pubic hair and adult breast contour.

On physical examination, a 3 x 4 cm swelling was noted in the right inguinal region, located medially above the mons pubis. The swelling was slightly tender, reducible, and displayed a cough impulse. Genital examination revealed normal labia majora and minora, but a dimple at the proposed vaginal opening, with no vaginal canal present. Other systemic examination, including cardiovascular, respiratory, and nervous system, were unremarkable. Based on the clinical findings, differential diagnosis of MRKH syndrome with inguinal hernia and complete androgen insensitivity with probable testicular swelling in the inguinal region was kept.

Ultrasound imaging of the abdomen and pelvis revealed that the right ovary was herniated into the inguinal canal and partially located within an intermuscular defect (Fig 1 A). The left ovary was normally positioned. The right ovary appeared slightly enlarged compared to the left. Additionally, the uterus and vagina were absent, and the right kidney was not detected. Magnetic resonance imaging (MRI) confirmed the findings (Fig. 1B,1C). The patient's karyotype was normal, 46XX, and the diagnosis of MRKH syndrome Type II was confirmed due to the presence of renal agenesis.

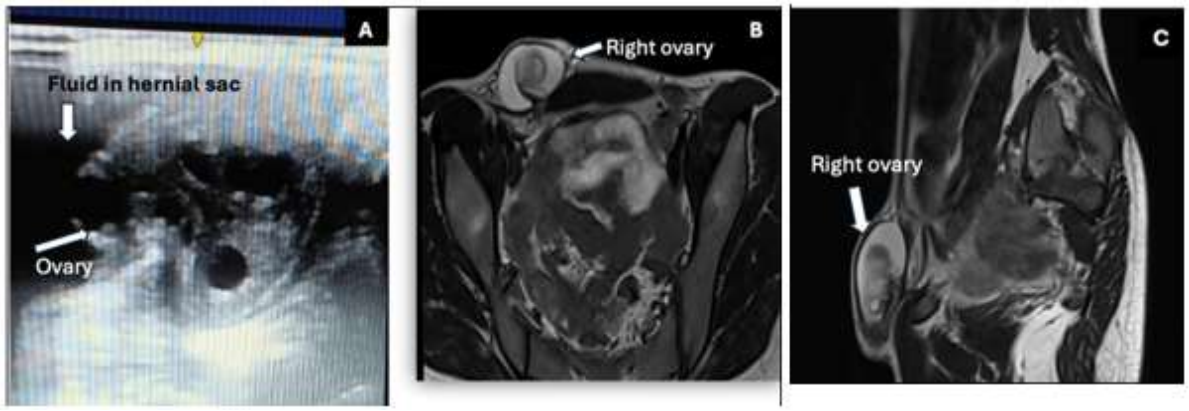

Figure 1: (a) Ultrasound image showing right ovary in hernial sac with fluid around it. (b) MRI image (trans axial section) showing ovary in inguinal canal. (c) MRI image (sagittal section) showing ovary in inguinal canal.

The patient underwent surgical exploration and repair of the inguinal hernia (fig. 2A, 2B). Upon exploration, a sliding-type right inguinal hernia was found (fig.2C), containing the right ovary, a rudimentary right-sided fallopian tube, and a rudimentary uterus (fig.2D, 2E). The herniated contents were reduced back into the pelvic cavity via the deep inguinal ring. The hernia defect was repaired with mesh placed over the deep inguinal ring and along the posterior wall of the canal (fig. 2F). The patient's postoperative recovery was uneventful.

Before discharge, the patient was counselled regarding potential future need for vaginoplasty and fertility management options like surrogacy.

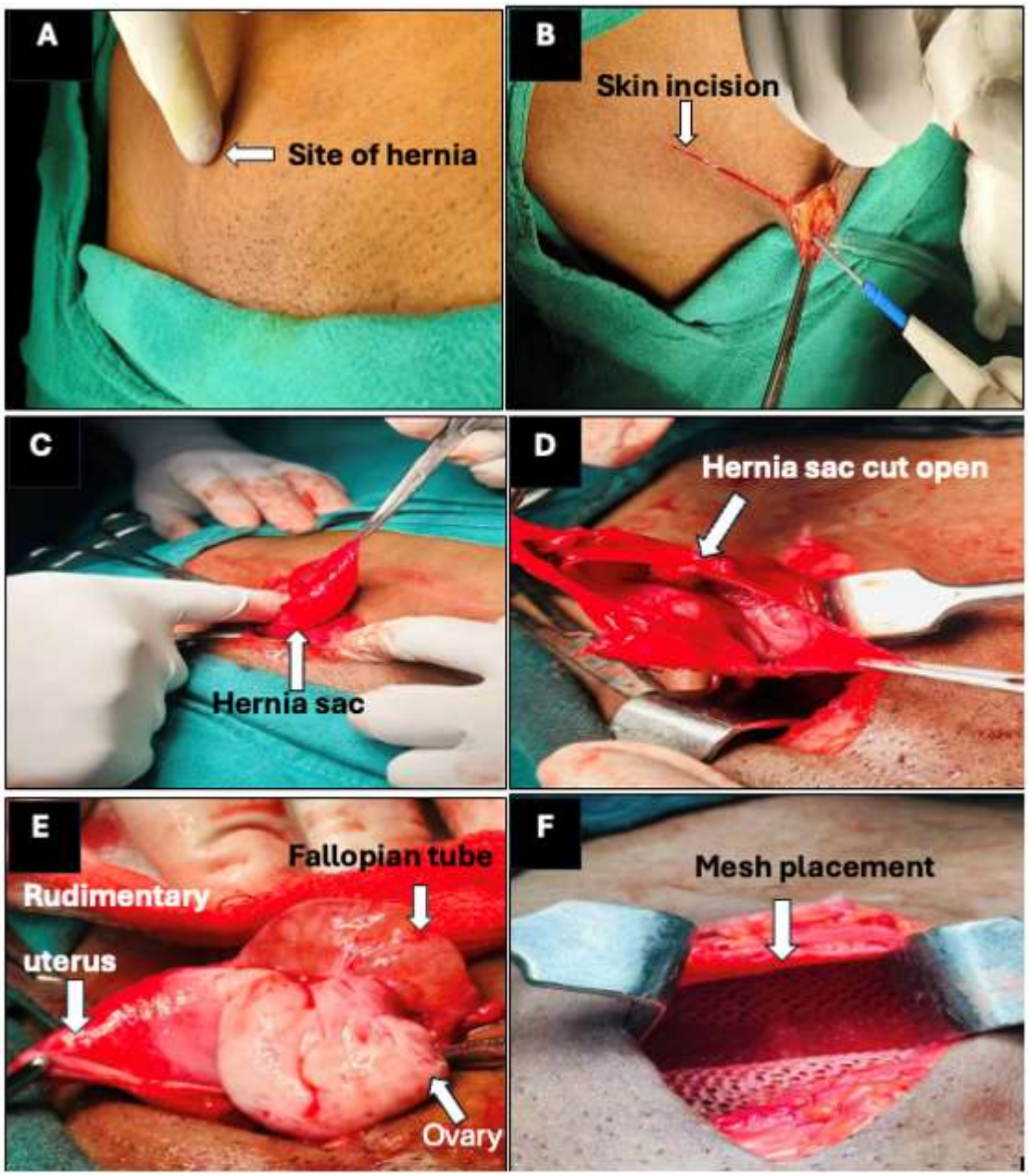

Figure 2: (a) Site of inguinal hernia, (b) Skin incision for hernia repair, (c) Hernia sac, (d) Hernia sac cut open, (e) Contents of the hernia rudimentary uterus, fallopian tube and ovary, (f) Placement of the mesh along posterior wall of inguinal canal and over the internal inguinal ring

Discussion

MRKH syndrome is a rare congenital condition, occurring in approximately 1 in 4,000 to 1 in 5,000 women, characterized by the absence or underdevelopment of the uterus and upper two-thirds of the vagina.[5] The ovaries typically function normally, and affected individuals present with primary amenorrhea despite normal secondary sexual characteristics, such as breast and pubic hair development. Karyotype is female 46 XX.

The management of MRKH syndrome is multifaceted, focusing on psychological, medical, and surgical interventions. Psychological counselling plays a crucial role in addressing the emotional and social impact of the condition. With appropriate care, individuals with MRKH can achieve healthy sexual relationships, and genetic offspring are possible through in vitro fertilization (IVF) and surrogacy.

The most common approach to vaginal reconstruction is non-surgical, involving daily self-dilation of the vaginal dimple. Studies by Jones et al. and Roberts et al. reported anatomical and functional success rates of 90–95% with this method.[8,9] For patients who do not respond to self-dilation, surgical creation of a neovagina by various techniques is considered a preferred treatment option. Ovarian herniation in MRKH also needs surgical correction in order to prevent torsion.

The inguinal canal is a narrow anatomical passage located along the lower part of the anterior abdominal wall. In females, it contains the round ligament of the uterus. Herniation of the ovary and other adnexal structures into the inguinal canal is rare and may occur due to a combination of factors. Several theories have been proposed to explain the occurrence of ovarian herniation. One theory, proposed by Thomson, suggests that non-fusion of the Mullerian ducts leads to increased ovarian mobility, predisposing the ovary to herniation.[10] Another theory, by Fowler, hypothesizes that congenital elongation of the ovarian ligaments contributes to ovarian herniation into the inguinal canal.[11] Additionally lengthening, of the broad ligament ovarian or suspensory ligaments due to multiparity may facilitate herniation.[12] MRI is the imaging modality of in MRKH to visualise status of uterus, ovaries and vagina.[13]

In MRKH syndrome, the absence of the uterus and vagina due to Mullerian agenesis does not preclude the risk of ovarian herniation. While rare, cases of ovarian herniation have been documented, and early diagnosis and treatment are critical to prevent complications such as irreducible ovary, torsion, infarction.[6] In cases of herniation, surgical relocation of the ovary and herniorrhaphy are the preferred treatments.[12-17]

Several case reports have highlighted the occurrence of ovarian herniation in patients with MRKH syndrome. In one retrospective study by Wang et al., MRI analysis of 83 patients with MRKH and 60 age-matched controls revealed a significantly higher incidence of abnormal ovarian position in MRKH patients.[18] A series of three cases described by Dai et al. involved young women with inguinal herniation of rudimentary uteri, one of which was an incarcerated hernia, all managed successfully with laparoscopic surgery.[19] Mohanty et al. described a case of bilateral ovarian herniation into the inguinal canal in a 20-year-old with MRKH syndrome,[6] while Al Omari et al. reported a case involving the herniation of the uterus, fallopian tubes, and ovaries in a 31-year-old woman with MRKH, successfully managed surgically.[20] Mahey et al described case of Mullerian agenesis and Turner mosaic syndrome with tubo-ovarian inguinal hernia in a 17 year old girl with left inguinal hernia and severe hypertension. Karyotype revealed Turner mosaic.[21]

Conclusion

Inguinal herniation of the ovary is a rare but important clinical finding in patients with MRKH syndrome and should be suspected in a patient diagnosed with this entity and coming with the complaint of swelling and pain in inguinal region. Early recognition and management are essential to avoid complications. Surgical repair is the treatment of choice, and counselling regarding future fertility and reconstructive options, such as vaginoplasty, should be provided in a manner sensitive to the patient's cultural and personal beliefs. As ovarian herniation in MRKH syndrome is likely under-reported, clinicians should maintain a high index of suspicion in patients presenting with inguinal hernias and primary amenorrhea.

References

1. Morcel K, Camborieux L, Guerrier D; Programme de Recherches sur les Aplasies Müllériennes. Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome. Orphanet J Rare Dis 2007 Mar;2:13.

2. Timmreck LS, Reindollar RH. Contemporary issues in primary amenorrhea. Obstet Gynecol Clin North Am 2003 Jun;30(2):287-302.

3. Saini R, Bains L, Kaur T, Lal P, Pal V, Beg MY, et al. Ovarian inguinal hernia - a possibility in MURCS syndrome. J Ovarian Res 2021 Sep;14(1):114.

4. Jafari R, Javanbakht M, Dehghanpoor F. Inguinal herniation of left ovary, fallopian tube and rudimentary left horn of bicornuate uterus associated with type 2 Mayer-Rakitansky-Kuster-Hauser (MRKH) syndrome in a teenage girl: A case report and literature review. Eur J Radiol Open 2020 Jan;7:100215.

5. Mohanty HS, Shirodkar K, Patil AR, Rojed N, Mallarajapatna G, Nandikoor S. A rare case of adult ovarian hernia in MRKH syndrome. BJR Case Rep 2017 May;3(3):20160080.

6. Daib A, Boughdir M, Ben Abdallah R, Hellal Y, Ben Malek MR, Gharbi Y, et al. A rare case of inguinal utero-ovarian hernia in girls. J. Clin. Med. Case Stud. 2017;2(2):1-3.

https://www.google.co.in/url?sa=t&source=web&rct=j&opi=89978449&url=https://www.rroij.com/open-access/a-rare-case-of-inguinal-uteroovarian-hernia-in-girls-.pdf&ved=2ahUKEwjbuu7syMyIAxUQzzgGHRlZCPUQFnoECCAQAQ&usg=AOvVaw08__ewx6Nhq28IuQJRKvB7.

7. Herlin M, Bjørn AM, Rasmussen M, Trolle B, Petersen MB. Prevalence and patient characteristics of Mayer-Rokitansky-Küster-Hauser syndrome: a nationwide registry-based study. Hum Reprod 2016 Oct;31(10):2384-2390.

8. Jones HW, Rock JA. Te Linde s operative gynecology. Lippincott Williams & Wilkins; 2015; 10.

9. Roberts CP, Haber MJ, Rock JA. Vaginal creation for müllerian agenesis. Am J Obstet Gynecol 2001; 185: 1349 53.

10. Thomson GR. Complete congenital absence of the vagina associated with bilateral herniae of uterus, tubes, and ovaries. Br J Surg 1948 Jul;36(141):99.

11. Fowler CL. Sliding indirect hernia containing both ovaries. J Pediatr Surg 2005 Sep;40(9):e13-e14.

12. Okada T, Sasaki S, Honda S, Miyagi H, Minato M, Todo S. Irreducible indirect inguinal hernia containing uterus, ovaries, and Fallopian tubes. Hernia 2012 Aug;16(4):471-473.

13. Kara T, Acu B, Beyhan M, Gökçe E. MRI in the diagnosis of Mayer-Rokitansky-Kuster-Hauser syndrome. Diagn Interv Radiol 2013;19(3):227-232.

14. Kumar A, Kumar A, Anwer M, Kumar D. Case of mullerian agenesis presenting as bilateral inguinal hernia with left sided irreducibility in a 21 years old female: A rare case report. Int J Surg Case Rep 2023 Feb;103:107895.

15. Al Omari W, Hashimi H, Al Bassam MK. Inguinal uterus, fallopian tube, and ovary associated with adult Mayer-Rokitansky-Küster-Hauser syndrome. Fertil Steril 2011 Mar;95(3):1119.e1-1119.e4.

16. Khan WF, Rathore YS, Pol MM, Singh G. Inguinal hernia with ovary as content-laparoscopic repair in a Mayer–Rokitansky– Küster–Hauser syndrome patient: case report. Int Surg J 2019;6(4):1421-1423.

17. Masoumi Shahrbabak M, Ebrahimi Meimand F. A rare case of Mayer-Rokitansky-Küster-Hauser syndrome with right ovarian torsion and hypoplasia of the left adnexa. J Surg Case Rep 2021 May;2021(5):rjab130.

18. Wang Y, Lu J, Zhu L, Chen R, Jiang B, Hou B, et al. Increased incidence of abnormally located ovary in patients with Mayer-Rokitansky-Küster-Hauser syndrome: a retrospective analysis with magnetic resonance imaging. Abdom Radiol (NY) 2018 Nov;43(11):3142-3146.

19. Dai Y, Qin C, Zhu L, Luo G. Hernia uterine inguinale associated with Mayer-Rokitansky-Küster-Hauser syndrome: Three case reports and literature review. Medicine (Baltimore) 2023 Feb;102(5):e32802.

20. Al Omari W, Hashimi H, Al Bassam MK. Inguinal uterus, fallopian tube, and ovary associated with adult Mayer-Rokitansky-Küster-Hauser syndrome. Fertil Steril 2011 Mar;95(3):1119.e1-1119.e4.

21. Mahey R, Rana A, Cheluvaraju R, Vyas S, Raj R, Bhatla N. An unusual association of type II Mayer-Rokitansky-Kuster-Hauser syndrome, turner mosaic syndrome and tubo-ovarian inguinal hernia- case report and review of literature. J Ovarian Res 2023 Feb;16(1):43.