Utero-Ovarian Inguinal Hernia in a Young Female with Mayer–Rokitansky–Küster–Hauser Syndrome Type 2

Dear Editor,
An inguinal hernial sac containing ovary, fallopian tube, and uterus associated with Müllerian dysgenesis, also known as Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome, is a very rare combination. MRKH syndrome has a reported incidence of 1 in 4500 newborn females. The transmission of this syndrome follows an autosomal-dominant pattern with variable expressivity and incomplete penetrance. Here, we describe a case of a young female who presented with primary amenorrhea and a right-sided irreducible inguinal swelling, which was later diagnosed as MRKH syndrome type 2 with utero-ovarian inguinal hernia. This case would increase the awareness among the surgeons about this rare condition, as early recognition and intervention can preserve the ovarian function in such patients.

A 26-year-old female presented to the outpatient general surgical clinic after referral from general practice. She had been experiencing a right-sided inguinal swelling since childhood. This swelling had progressively worsened in size in the past 10 years and had begun causing symptomatic pain. The swelling was initially reducible, but become irreducible a few years ago. The patient revealed she had primary amenorrhea. Secondary sexual characteristics were well developed. She had normal stature including height for age and there was no hearing abnormality. She had a blind-ending vagina on gynecological examination. Differential diagnoses included imperforate hymen, androgen insensitivity syndrome, MRKH syndrome, or true hermaphrodite. Ultrasound pelvis revealed absent left ovary and gonadal structure/ovary in the right inguinal sac having mixed echogenicity but no fluid. Computerized tomography scan showed mild cardiomegaly, agenesis of the right kidney, and absent uterus, with a hernial sac containing gonadal soft tissues [Figure 1]. Magnetic resonance imaging (MRI) of the pelvis confirmed the same findings; besides, there were hyperintense signals in the sac [Figure 2]. Echocardiography showed mitral stenosis with moderate tricuspid regurgitation. Laboratory investigations showed testosterone, 1.15 nmol/L; FSH, 7.46; and LH, 9 mIU/ml. Karyotyping of this patient was that of a normal female (i.e., 46XX). Diagnostic laparoscopy was done, which showed absent left ovary and uterus and right-sided indirect inguinal hernia. An inguinal incision was given, contents of sac were rudimentary uterus with ligaments and right fallopian tubule and ovary [Figure 3]. Wedge biopsy of the ovary was taken, and structures were repositioned back in the pelvic cavity. Lichtenstein’s repair was done. Histopathology report confirmed ovarian tissue. The patient was discharged and referred to a gynecologist for further follow-up.
Müllerian dysgenesis is the failure of the Müllerian duct to develop during the embryonic period. This can result in maldevelopment of the uterus, upper two-thirds of the vagina, or both as these have a common paramesonephric ductal origin. Ovaries remain unaffected, as their embryonic development is distinct from these.[2] This occurrence in isolation is less frequent than cases where Müllerian dysgenesis is found to coexist with any of the MURCS associations (Müllerian duct aplasia, renal dysplasia, and cervical somite anomalies). These are termed as MRKH type 2 syndromes.[3] During the embryonic period, the vaginal process and the round ligament pass through the inguinal canal. If vaginal process remains partially patent in female, any intrabdominal structures including ovaries, fallopian tubes, and sometimes uterus can herniate as inguinal hernia. Malformation of suspensory ligament is attributed to the uterine herniation in inguinal hernia.[4]

MRKH syndrome is the second most prevalent disorder among the most common causes of primary amenorrhea. On physical examinations, such patients have normal body hair, height, and external genitals. They have normal female hormones levels and genotype.[5]

In 2011, Al Omari et al.[6] reported a case of utero-ovarian inguinal in MRKH syndrome and stated that only two similar cases were reported in the literature, and that their patient was the third. In 2018, Verma et al.[7] reported a case of MRKH type 2 with a hernial sac containing rudimentary uterus and ovary.

MRI is an investigation of choice, as it helps in identifying absent or rudimentary uterus and other associated anomalies. Management of this rare condition involves reducing sac content, especially ovary, back into its position to preserve its function, followed by hernial repair. A detailed history and pelvic examination should be carried out prior to surgery in females with inguinal hernia, as there is an association of developmental anomaly in whom a hernial sac could possibly contain ovary or fallopian tubule. A detailed history, pelvic examination and work-up (including hormones levels, ultrasound, MRI and karyotyping) should be carried out prior to surgery in females with inguinal hernia, especially when there is primary amenorrhea, as there is an association of developmental anomaly in whom a hernial sac could possibly contain ovary or fallopian tubule.[7,8]

**Declaration of patient consent**

The authors certify that they have obtained all appropriate consent forms from the patient. The patient has given her consent for images and other clinical information to be reported in the Journal. The patient understands that her name and initials will not be published, and due efforts will be made to conceal the identity.

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**Conflicts of interest**

There are no conflicts of interest.

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**REFERENCES**

1. Verma R, Shah R, Anand S, Vaja C, Gaikwad K. Mayer-Rokitansky-Küster-Hauser Syndrome presenting as irreducible inguinal hernia. Indian J Surg, 2018;80:93-5.
2. Kamio M, Nagata T, Yamasaki H, Yoshinaga M, Douchi T. Inguinal hernia containing functioning, rudimentary uterine horn and endometriosis. Obster Gynecol. 2009;113:563-6.
3. Morcel K, Camborieux L, Programme de Recherches sur les Aplasies Müllerennes; Guerrier D. Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome. Orphanet J Rare Dis. 2007;2:13.
4. George EK, Oudesluys-Murphy AM, Madern GC, Cleyndert P, Blomjous JG. Inguinal hernias containing the uterus, fallopian tube, and ovary in premature female infants. J Pediatr. 2000;136:696-9.
5. Kriplani A, Banerjee N, Aminni AC, Kucheria K, Takkar D. H Hernia uterus inguinal in a 46, XX female. A case report. J Reprod Med. 2000;45:48-50.
6. Al Omari W, Hashimi H, Al Bassam MK. Inguinal uterus, fallopian tube, and ovary associated with adult Mayer-Rokitansky-Küster-Hauser syndrome. Fertility and Sterility. 2011;95:1119.e1-e4.
7. Bazi T, Berjawi G, Scound M. Inguinal ovaries associated with müllerian agenesis: Case report and review. Fertil Steril. 2006;85:1510.e5-8.
8. Bidus MA, Martin [N], Magann EE. Mayer-Rokitansky-Küster-Hauser Syndrome Presenting as an Inguinal Mass and Hernia in the Female Patient. Female Pelvic Med Reconstr Surg. 2004;10:193-5.

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