CASE REPORT

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A Case Report of Type-II Mayer-Rokitansky-Küster-Hauser Syndrome with Ovarian Inguinal Hernia Presenting with an Adnexal Mass

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ABSTRACT Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a rare congenital disorder of the genitourinary tract seen in pediatric females. It is of 2 types. Type I has mullerian anomalies while Type II has associated anomalies. It may be associated with an inguinal hernia. The contents of the hernial sac are mostly the omentum or small intestine. Rarely, unusual contents like ovary and fallopian tubes may also be found in the hernia. The association of MRKH syndrome with ovarian hernia has been reported in a few cases in the literature. Our case is a similar rare case of a pediatric female of MRKH syndrome Type II with an inguinal hernia having an ovary as its content who presented with an adnexal mass.

Keywords: Mayer-Rokitansky-Küster-Hauser syndrome; inguinal hernia; adnexal mass

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a rare congenital disorder of the genitourinary tract seen in pediatric females. The incidence reported is 1 in 5,000 live female births.[1] It is associated with mullerian aplasia with or without other associated anomalies. It is of two types. Type I where hypoplasia/agenesis of the upper part of vagina and uterus is found and Type II has associated renal, vertebral, skeletal, and cardiac malformations.[2] These patients have a female karyotype and normal secondary sexual characters. Inguinal hernia which is the most common hernia in adults but uncommon in females may be seen along with this syndrome. The contents of the hernial sac are the omentum or small intestine but the appendix, caecum, sigmoid colon, and urinary bladder may also be found.[3] A few cases have been reported having unusual contents in hernial sacs such as ovaries and fallopian tubes and the incidence reported is 2.9%.[4] Prodromidou et al. did a systematic search to know the existing evidence on ovarian inguinal hernia and retrieved 17 such cases.[5] The most common presentation of MRKH syndrome is primary amenorrhoea but manifestations due to associated anomalies may be seen. We hereby report a rare case of Type II MRKH syndrome with an uncomplicated indirect inguinal hernia having ovary as its content with a pelvic mass in a pediatric female.

CASE REPORT

A 12-year-old female, moderately built with average height presented in the outpatient department with complaints of dull pain in the lower abdomen and abdominal distension for the past one month. Her bladder and bowel habits were normal. She had not attained her menarche. All the secondary sexual characters were well developed. General physical examination was normal. Her 4 sibling sisters had timed and regular menstruation.

On abdominal examination, a 3x3 cm painless lump was palpable in the left inguinal region (Figure 1). On local examination, vagina was absent, instead, a dimple was seen. A firm, cystic, non-tender mass of 8x8 cm was felt on rectal examination.

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Ultrasonography of the abdomen and pelvis showed the absence of uterus and the left kidney. Left ovary measured 33x28 mm and was lying in the subcutaneous plane in left inguinal region. A solid cystic mass, measuring 90x82x60 mm was seen in the pelvis. On color Doppler, there was no tortuosity in the pedicle (Figure 2).

Magnetic resonance imaging (MRI) of the abdomen and pelvis confirmed the absence of the uterus and the left kidney and ectopic location of the left ovary. A well-defined peripherally solid and centrally cystic lesion measuring 90x90x60 mm was noted in the pelvis. Computed tomography intravenous pyelography (CTIVP) confirmed absent left kidney (Figure 3). The right kidney was 96x60 mm in size, malrotated with hilum facing posteromedially. Figure 4 shows an axial section imaging of CTIVP showing the ectopic location of the left ovary in the subcutaneous plane in the inguinal region. The presence of bony defect with the bifid spinous process was also noted involving sacral vertebrae with open spinal canal suggestive of spina bifida. Neurosurgeon consultation was taken for the incidental diagnosis of spina bifida and since she was asymptomatic for the same with no spinal cord anomaly, no active intervention was advised. Carcinoembryonic antigen, alpha-fetoprotein, beta human chorionic gonadotropin, and lactate dehydrogenase were within normal limits. Cancer antigen 19-9 and cancer antigen 125 were 49.81 U/mL and 49.0 U/mL, respectively. Karyotyping was done to rule out the possibility of dysgenetic gonads. She had a normal female genotype of 46XX.

Counseling of the parents was done regarding the presence of congenital anomaly, ovarian hernia, and the need of performing unilateral/bilateral oophorectomy in case of infarcted ovary due to torsion or any malignancy detected in the frozen section.

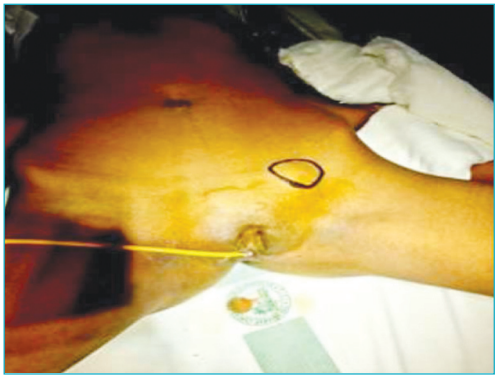

FIGURE 1: Painless palpable lump in the left inguinal region.

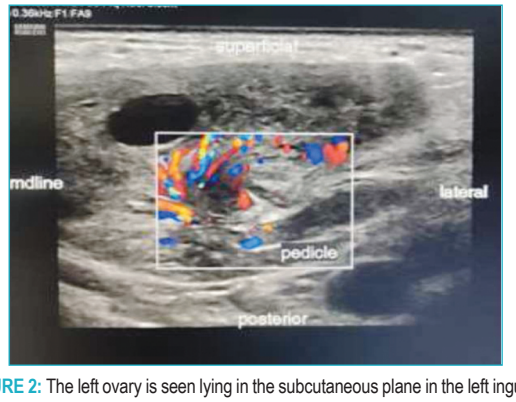

FIGURE 2: The left ovary is seen lying in the subcutaneous plane in the left inguinal region with its vascular pedicle. On examination with colour Doppler, there was no tortuosity in the pedicle.

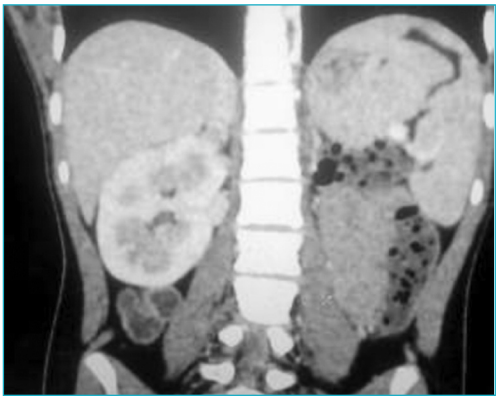

FIGURE 3: A coronal section of computed tomography intravenous pyelography showing absent left kidney.

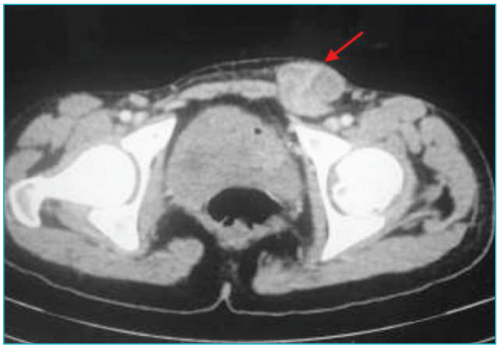

FIGURE 4: An axial section imaging of computed tomography intravenous pyelography showing ectopic location of left ovary in the subcutaneous plane in inguinal region (marked with arrow).

The patient underwent staging laparotomy. Intraoperatively, a 6x7 cm right ovarian mass was seen. Systematic exploration of the abdomen was normal except for an empty left renal fossa. Uterus was rudimentary and a 1 cm thin sheet-like uterine tissue was seen in the place of the uterus. The left infundibulopelvic ligament was seen protruding through the inguinal canal into the subcutaneous plane (Figure 5). Cystectomy could not be done due to dense adhesions with adjacent bowel loops. Thus, mass along with right fallopian tube was removed and sent for frozen section examination. The frozen section showed a benign hemorrhagic cyst.

A mediolateral incision was given over the palpable lump. The ovary was seen lying in the subcutaneous plane. Hernial contents were reduced and the left fallopian tube and the left ovary were brought back into the pelvis to their normal positions and were found to be normal looking. Since the ability to carry a pregnancy in her uterus was not possible, left salpingectomy was done. Ovariopexy to the left lateral pelvic wall was done followed by inguinal hernia repair. Final histopathology was benign and peritoneal cytology was also negative for malignant cells. At 3 months postoperatively, on a follow-up ultrasonography of the pelvis, the left ovary was normal looking measuring 49x39 mm, and serum follicle stimulating hormone and serum luteinizing hormone levels were 1.83 mIU/mL and 1.37 mIU/mL, respectively.

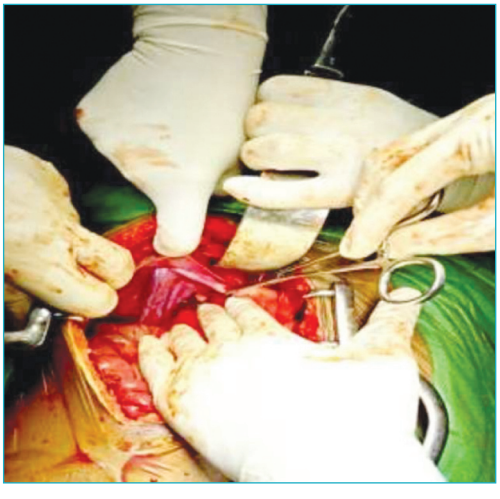

FIGURE 5: Left infundibulopelvic ligament was seen protruding through the inguinal canal into subcutaneous plane.

Informed consent was taken beforehand to publish this case because of its rarity and the identity.

DISCUSSION

Association of ovarian hernia in MRKH syndrome has often been attributed to congenital anatomical defects the mechanism of which is not clear. One such explanation is persistent processus vaginalis in female infants which is an outpouching of the parietal peritoneum through the inguinal canal associated with the round ligament known as the canal of Nuck.[4,6-8] In a hypothesis by Thomson, when the mullerian ducts fail to fuse, ovaries become excessively mobile causing protrusion of ovaries, fallopian tubes, and even the entire uterus into the hernial sac.[9] In another theory proposed by Fowler, elongated ovarian suspensory ligaments may be the primary etiology or the secondary effect of a hernia.[10]

As the secondary sexual characters in MRKH syndrome are normal, presentation is usually with complaints of delayed menarche. However, a diagnosis can be made on imaging done for other problems. In our case, multiple imaging studies were necessary to assist in diagnosis, including ultrasound, MRI, and CTIVP. Ovaries, when trapped in the inguinal region, can lead to complications like torsion, irreducibility, salpingitis, gangrene, etc. thus a delayed diagnosis might result in loss of endocrine function of the ovary and future infertility. In such patients, thorough counseling of both the parents is required. Apart from management of anomaly, appropriate surgical management is required to ensure the preservation of ovaries.

Hence, awareness of the possibility of ovarian hernia in patients of MRKH, prompt diagnosis, and early intervention to salvage ovaries is of utmost importance. A multidisciplinary approach including a radiologist, a gynaecologist having expertise in pelvic anatomy and fertility management is vital for management. Also, the need for a counselor cannot be overlooked as the presence of mullerian anomaly can have a psychological impact.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Pariseema Dave;* ***Design:*** *Bijal Patel;* ***Control/Supervision:*** *Pariseema Dave;* ***Data Collection and/or Processing:*** *Nishu Dahingra;* ***Analysis and/or Interpretation:*** *Bijal Patel;* ***Literature Review:*** *Nishu Dahingra;* ***Writing the Article:*** *Nishu Dahingra;* ***Critical Review:*** *Pariseema Dave;* ***References and Fundings:*** *Bijal Patel;* ***Materials:*** *Nishu Dahingra.*

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