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Inguinal Ovarian Herniation: An Unusual Presentation of Mayer-Rokitansky-Küster-Hauser Syndrome Type B: A Case Report

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ABSTRACT

Inguinal ovarian herniation is an uncommon finding in adult women and may be associated with congenital Müllerian anomalies such as Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome. We report the case of a 37-year-old woman presenting with primary amenorrhea. Clinical examination revealed a short blind-ending vagina. Pelvic MRI demonstrated complete uterovaginal agenesis associated with a solitary right kidney and a left inguinal hernia containing a hypoplastic ovary. These findings were consistent with MRKH syndrome type B. This case emphasizes the diagnostic value of MRI in detecting ectopic ovarian localization and associated congenital anomalies in patients with primary amenorrhea.

KEYWORDS

Ovarian inguinal hernia, MRKH syndrome type B, Primary amenorrhea, Ectopic ovary.

MAIN ARTICLE

INTRODUCTION

Inguinal herniation of the ovary is a rare condition in adult females and is more frequently encountered in pediatric populations. In women, ovarian ectopia may occur in association with congenital anomalies of Müllerian duct development, particularly Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome.

MRKH syndrome is characterized by congenital agenesis of the uterus and upper two-thirds of the vagina in phenotypically normal females with normal ovarian function and karyotype. The syndrome is divided into:

- **Type A:** isolated uterovaginal agenesis.
- **Type B:** uterovaginal agenesis associated with extragenital malformations, especially renal and skeletal anomalies.

The coexistence of ovarian inguinal herniation and MRKH syndrome is uncommon and clinically significant because of the potential risk of ovarian torsion, infertility issues, and diagnostic delay [1,2].

CLINICAL PRESENTATION

A 37-year-old woman presented with primary amenorrhea. Clinical examination revealed a short blind-ending vagina measuring approximately 2 cm.

Initial ultrasound examination demonstrated absence of the uterus and vagina associated with a solitary right kidney, raising suspicion for MRKH syndrome.

MRI Findings

- Complete absence of the uterus and vagina, consistent with uterovaginal agenesis. (Figure 1)
- A normal right ovary with preserved follicular activity and homogeneous signal intensity, measuring 28 × 13 mm. (Figure 3)
- A left inguinal hernia containing the left ovary, which appeared hypoplastic and follicular, measuring 16 × 8 mm. (Figure 2)

- A small adjacent fluid collection surrounding the herniated ovary. (Figure 2)
- No evidence of adnexal torsion or ischemic complication.
- Empty left renal fossa consistent with left renal agenesis. (Figure 3)

These findings supported the diagnosis of MRKH syndrome type B.

DISCUSSION

Ovarian inguinal herniation results from protrusion of the ovary through the inguinal canal. In females, this condition is rare and often associated with congenital anomalies involving abnormal ligamentous support or Müllerian duct malformations [2,3].

In patients with MRKH syndrome, defective Müllerian development may alter ovarian positioning and predispose to ovarian ectopia or herniation.

Clinical Presentation

Patients may present with:

- Inguinal swelling
- Pelvic pain
- Primary amenorrhea
- Infertility
- Incidental imaging findings

In this case, primary amenorrhea was the presenting symptom, while the ovarian herniation was identified on MRI.

Radiological Features [3]

MRI is the gold standard imaging modality for evaluating Müllerian anomalies and ectopic ovaries because of its excellent soft tissue contrast and multiplanar capability.

Characteristic findings include:

- Absence of the uterus and upper vagina
- Normal or ectopic ovaries with preserved follicles

- Associated renal anomalies
- Identification of inguinal ovarian localization

The presence of follicles within the inguinal lesion strongly supports ovarian tissue identification.

Associated Anomalies

MRKH syndrome type B is frequently associated with:

- Renal agenesis or ectopia
- Skeletal anomalies
- Ovarian malposition
- Hearing abnormalities [3,4].

Renal anomalies are the most commonly associated malformations.

Differential Diagnosis

Differential diagnoses of inguinal masses in women include:

- Inguinal lymphadenopathy
- Canal of Nuck cyst
- Inguinal endometriosis
- Lipoma
- Herniated bowel loops

MRI allows accurate tissue characterization and diagnosis [3, 4].

Management

Management depends on symptoms, ovarian viability, and reproductive considerations.

Treatment options include:

- Surgical reduction and hernia repair
- Oophoropexy to preserve ovarian function
- Vaginal reconstruction in MRKH syndrome

- Fertility counseling and assisted reproductive options

Early diagnosis is important to prevent ovarian torsion and preserve reproductive potential [3,5].

CONCLUSION

This case illustrates a rare presentation of ovarian inguinal herniation associated with MRKH syndrome type B in an adult patient presenting with primary amenorrhea. MRI played an important role in diagnosing uterovaginal agenesis, identifying the ectopic ovary, and detecting associated renal agenesis. Recognition of ovarian herniation in MRKH syndrome is essential because of its therapeutic and reproductive implications.

FIGURES

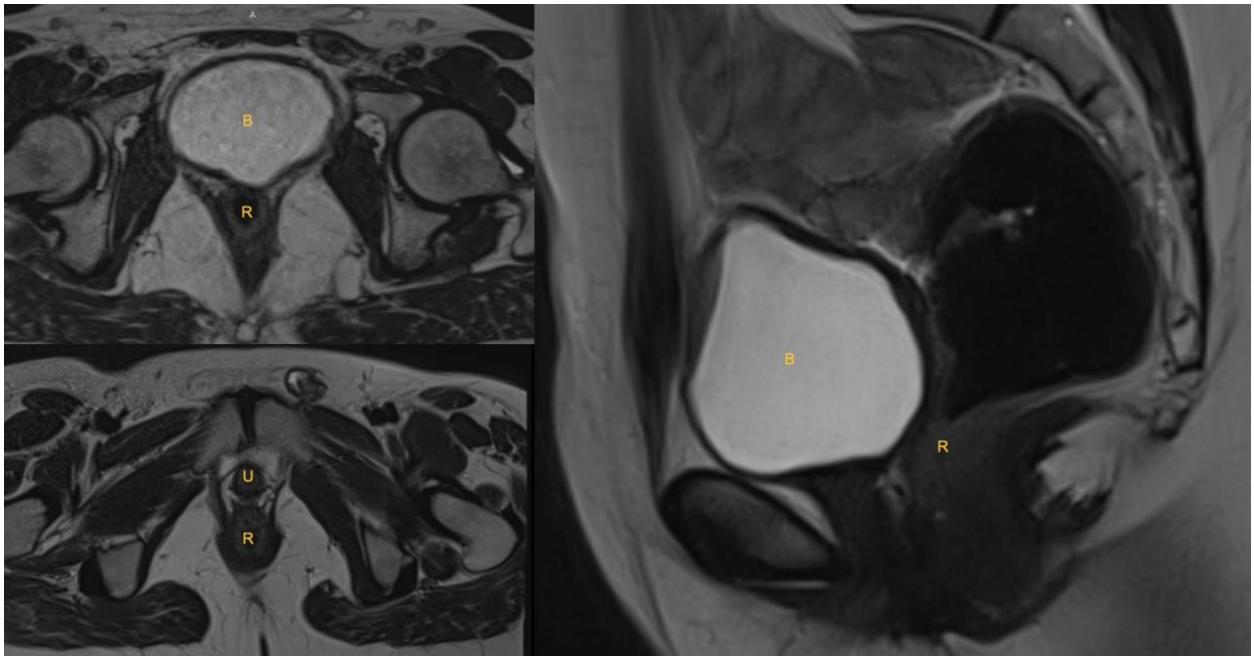

Figure 1 : Pelvic MRI in axial and sagittal T2 weighted sequences demonstrating complete uterovaginal agenesis.

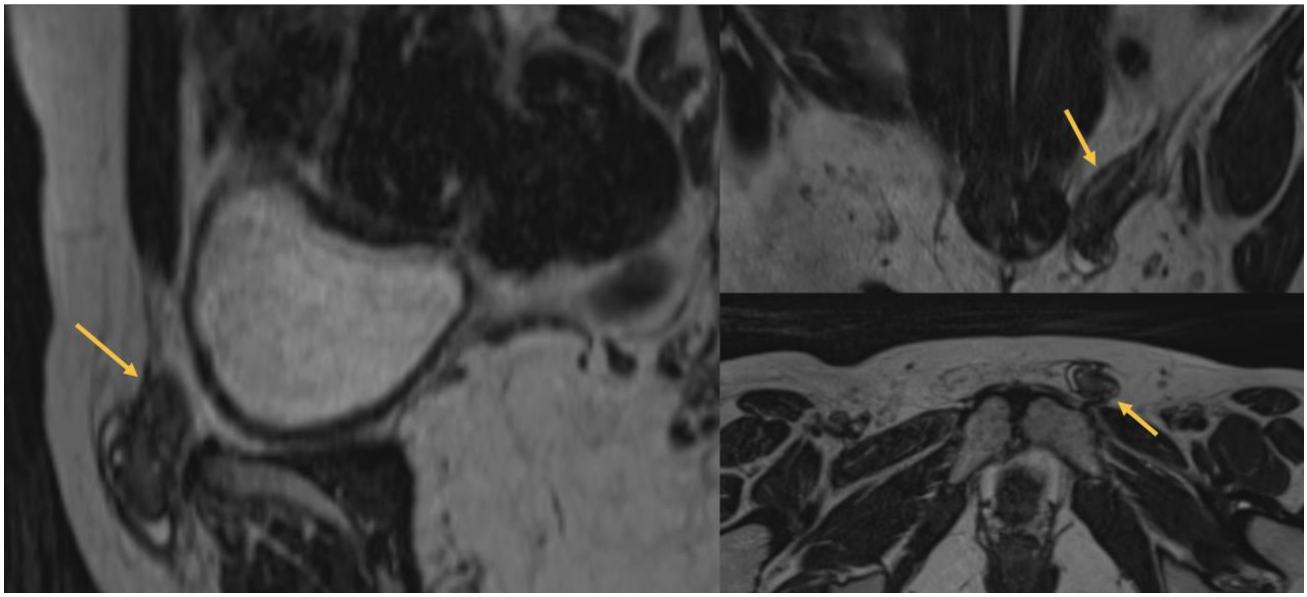

Figure 2 : T2 weighed sequences in axial coronal and sagittal view showing left inguinal hernia containing a hypoplastic follicular ovary with adjacent fluid collection.

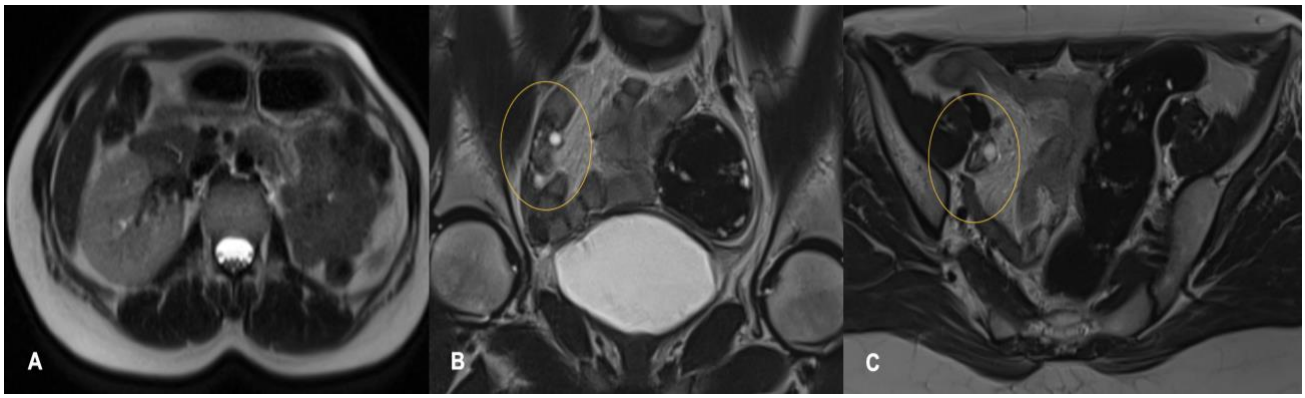

Figure 3: T2 weighed sequences in axial and coronal view showing a left kidney agenesis (A) and a normal placed follicular right ovary (B and C).

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