Case 16232

Mrkh with right ovarian inguinal hernia
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Section: Genital (female) imaging
Area of Interest: Abdomen
Procedure: Diagnostic procedure
Procedure: Complications
Imaging Technique: MR
Special Focus: Congenital Hernia Case Type: Clinical Cases
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Patient: 20 years, female

Clinical History:
A 20-year-old female presented to department of gynecology with complaints of primary amenorrhoea and pain in right inguinal region. On examination, secondary sexual characteristics were normal. A painful swelling was noted in right inguinal region. Patient had undergone gynaecologic US outside, for which the documents were not available. MRI was done.

Imaging Findings:
On MRI, uterus was not visualized. A nodular lesion with lobulated margin was seen in left adnexal region measuring about 17 x 13 mm with STIR hyperintensity, T2 and T1 isointensity to muscle, thas was interpreted as possible rudimentary horn/hypoplastic Mullerian bud. No T2 hyperintense strip/hemorrhagic component was seen within nodular lesion - possibly no functional endometrium. Upper 2/3rd of vagina was not visualized. The left ovary was normally visualized (30 x 28 mm). Right inguinal hernia was seen with partial extension of right ovary within the hernial sac. Right ovary was bulky. STIR hyetogenous and T2 hyperintense signal with hypointense areas within - likely due to hemorrhage/congestion. Right ovary measured 66 x 28 mm.

Discussion:
Background: Mayer-Rokitansky-Kuster-Hauser syndrome (MRKHS) is a rare congenital Müllerian anomaly, which typical form is characterized by partial or complete absence of the uterus with an absent or hypoplastic vagina. It was first described by Mayer (1829) and Rokitansky (1838) as agenesis of the uterus and vagina, due to abnormal development of the uterine ducts [1].
As the secondary sexual characters are normal, these patients are not diagnosed until puberty. However a diagnosis might be made on imaging for other health problems.
MRKHS has two subtypes: the typical (also called Rokitansky sequence, type I, type A or isolated) and the atypical form (type II or type B) [2].
In patients with typical form, the only affected part is the caudal part of the Müllerian duct. The atypical form of MRKHS is associated with other anomalies, including mainly renal anomalies such as unilateral agenesis, ectopia of one or both kidneys, or horseshoe kidney [3].
Inguinal hernia is the most common hernia occurring in adults, more commonly in males. Females are less affected, most commonly presenting in late adulthood.
Mostly inguinal hernial sac contains omentum or small bowel, but caecum, appendix and sigmoid colon are seen at
times and urinary bladder may also protrude as content [4].
Inguinal hernias containing ovaries (ovarian hernias) occur very rarely in adult females (approximately <3%) [5].
Only few cases have been reported of MRKH with inguinal hernia.

Clinical Perspective: young females with primary ammenorhea, usually the secondary sexual characters (external appearances) are normal. Inguinal hernias though rarely seen, must be evaluated for contents. Associated renal anomalies (eg. dysplasia, ectopia, horse-shoe kidney) and musculoskeletal anomalies (eg. scoliosis) should also be evaluated.

Imaging Perspective: MRI is the imaging modality of choice, showing absent uterus, and upper 2/3 of the vagina. Ovaries are placed normally (in this case right ovary is abnormally placed - in right inguinal region). Renal abnormalities are associated in many cases, so the imagins studies must include urinary system. This entity can show rudimentary uterine bud without or with functioning endometrium. If the latter is present can cause cyclic pain.

Outcome: ovarian torsion, ovarian necrosis, salpingistis are few complications of ovarian inguinal hernia. Patient underwent surgery for inguinal hernia. In this case the herniated ovary was incarcerated and showed hemorrhagic cysts in the biopsy sample. Patient was advised to undergo vaginoplasty.

Take Home Message, Teaching Points
In MRKHm, look for functioning endometrium, renal abnormalities and ovarian complications.

Written informed patient consent for publication has been obtained.

**Differential Diagnosis List:** MRKH with right inguinal hernia with necrosed ovary as its content., WNT4 syndrome - Biason-Lauber syndrome, Androgen insensitivity

**Final Diagnosis:** MRKH with right inguinal hernia with necrosed ovary as its content.

**References:**


Figure 1

Description: AXIAL STIR IMAGE SHOWING RIGHT OVARY IN RIGHT INGUINAL REGION (STAR)
Origin: Arora B, Department of Radiology, SDMH, Jaipur, Rajasthan, India

Description: AXIAL T1 FSE SHOWING RIGHT OVARY IN RIGHT INGUINAL REGION (STAR)
Origin: Arora B, Department of Radiology, SDMH, Jaipur, Rajasthan, India
Description: SAGITTAL T2 FSE SHOWING RIGHT OVARY IN RIGHT INGUINAL REGION (STAR)
Origin: Arora B, Department of Radiology, SDMH, Jaipur, Rajasthan, India
Figure 2

a

Description: AXIAL T1 FSE SHOWING RUDIMENTARY HORN (ARROW)
Origin: Arora B, Department of Radiology, SDMH, Jaipur, Rajasthan, India

b

Description: CORONAL T2 FRFSE SHOWS RUDIMENTARY HORN ON LEFT SIDE (ARROW)
Origin: Arora B, Department of Radiology, SDMH, Jaipur, Rajasthan, India
Description: CORONAL STIR SHOWS RUDIMENTARY HORN ON LEFT SIDE (ARROW)

Origin:
Arora B, Department of Radiology, SDMH, Jaipur, Rajasthan, India
Figure 3

a

Description: AXIAL STIR SHOWING LEFT OVARY IN NORMAL LOCATION
Origin: Arora B, Department of radiology, SDMH, Jaipur, Rajasthan, India

b

Description: sagittal T2 image showing absent uterus and upper 1/3 vagina
Origin: Arora B, Department of radiology, SDMH, Jaipur, Rajasthan, India