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A case of Mayer Rokitansky Kuster Hauser syndrome

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Abstract

Purpose & study object: We herewith present a case of unusual mullerian duct anomaly presenting with primary amenorrhea and some Past history suggestive of hymenotomy (details not available).

Introduction: Mullerian duct anomaly is approximately 6.7% in general population. Incidence is higher in infertility 7.3% and recurrent miscarriages 16%. We present here a case of MRKH syndrome, which is a Abnormality of development of the female genital tract: partial or complete absence (agenesis) of the uterus, absent or hypoplastic vagina, normal fallopian tubes, ovaries, normal external genitalia.

Material & Method: Basic investigation of USG was performed which revealed unicornuate uterus, elongated tapered structures suggestive of Rt. Hydrosalpinx and normal ovaries. To evaluate further communication between uterus and vagina, CT scan & MRI were performed. Graded C.T. cystogram also showed a false tract like structure going towards perineum. Since these findings were inadequate for further management and treatment – diagnostic laparoscopy and cystoscopy was performed.

Conclusion: There was unicornuate uterus with rt. tube showing hydrosalpinx with blood collection, absent cervix and absent proximal vagina. These findings were confirmed with help of diagnostic laparoscopy and cystoscopy.

Keywords: Mullerian Duct Anomaly, Mayer Rokitansky Kuster Hauser Syndrome

Introduction

Purpose: To Find out mullerian duct anomalies with help of usg, ct, mri and to establish a combination and to explain it on basis of embryology.

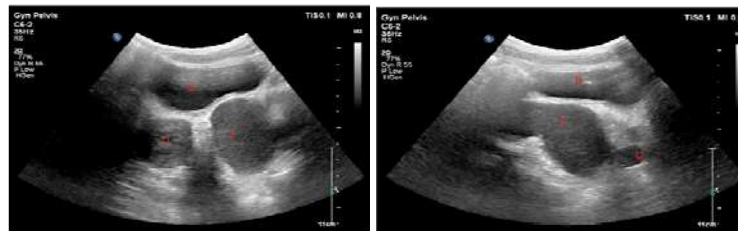
History: A 15 year old female came with chief complaints of Pain in lower abdomen, since 15 days

Hematuria on and off, since 5 days. Patient had Primary Amenorrhoea for which she underwent hymenotomy procedure 3 months back, she was advised ultrasonography for continued amenorrhoea.

Methods

1) Ultrasonography

Uterus -unicornuate with continuity missing from cervix, adjacent to it is a dilated tube like structure and above which bladder is seen. A well defined cystic lesion tapering towards fundus of uterus with normal right ovary seen separately-s/o Hydrosalpinx. Kidneys and other abdominal viscera normal.



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Ultrasonography revealed

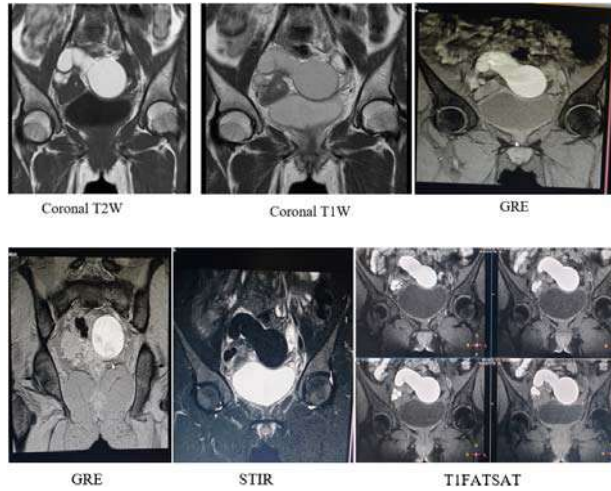
Uterus: mild heterogeneously echogenic collection in the endometrial cavity.

Enlarged tubular structure noted to the side of uterus with echogenic collection noted with in.

Bladder: slightly thick walled bladder with post void residue: Nil

Findings suggestive of hematometra with Hydrosalpinx
Further MRI PELVIS was advised.

MRI PELVIS



Unicornuate uterus with single tube with minimal collection in cavity. Rt. tube grossly dilated and filled up with hemorrhagic fluid. Single ovary is identified close to dilated tube. Doubtful fistulous tract between bladder base to perineum which do not fit in embryological basics.

On T1W: hyperintense signal of tube.

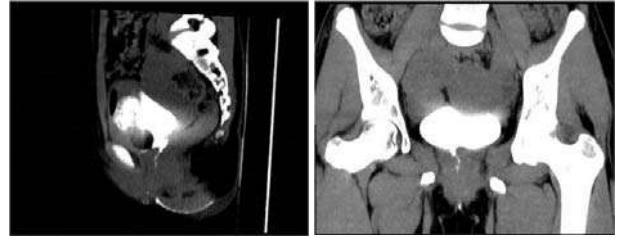
On T2W: hyperintense signal of tube.

On STIR: hypointense signals of tube.

On T1 FAT SAT: hyperintense signals of tube.

GRADED CT CYSTOGRAM was advised (in view of fistulous communication).

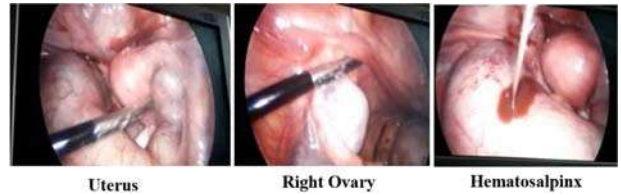
Graded Ct Cystogram



Graded CT cystogram reveals a fistulous communication between base of bladder and perineum

Results

- 1) It confirms unicornuate uterus.
- 2) Hydrosalpinx showing remnants of blood – Hematosalpinx,
- 3) Ovary on Left-slightly hypoplastic.
- 4) Absent communication between cervix and perineum –agenesis of vagina
- 5) Graded CT cystogram showed fistulous tract extending from trigone to perineum.
- 6) Diagnostic laparoscopy plus cystoscopy was advised.



Conclusion

Different imaging techniques revealed following findings

- 1) Unicornuate uterus.
- 2) Right. side hematosalpinx
- 3) Absent communication between cervix and perineum-agenesis of vagina indicating possibility of Mayer Rokitansky Kuster Hauser Syndrome (Type 1)

Discussion

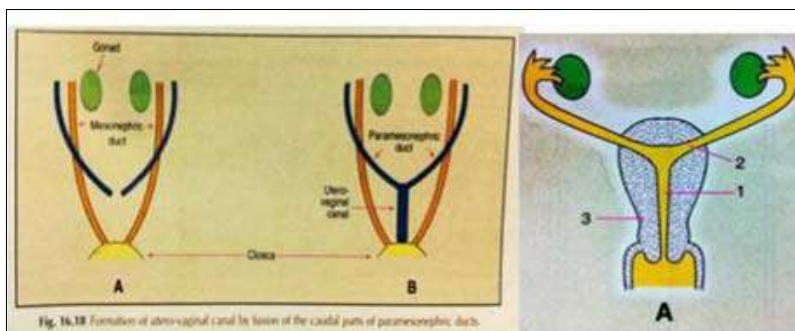
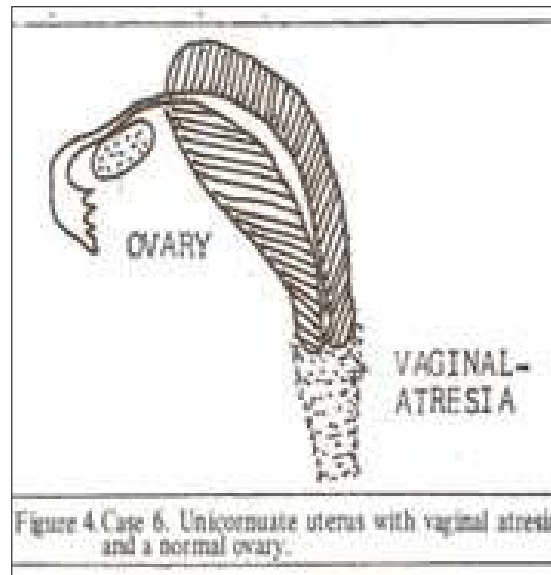
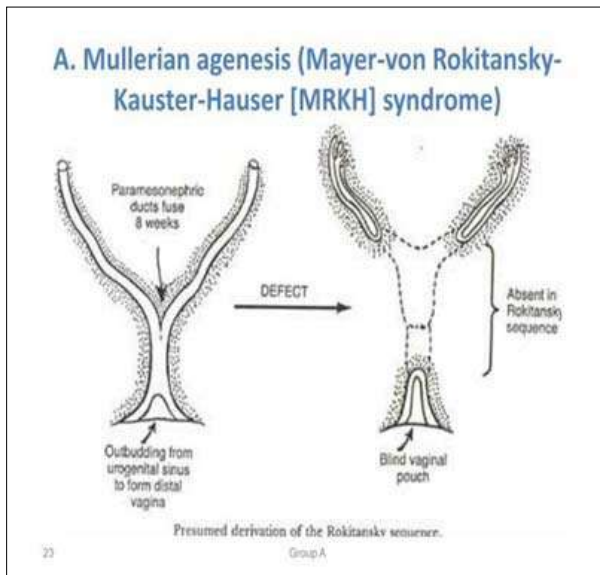
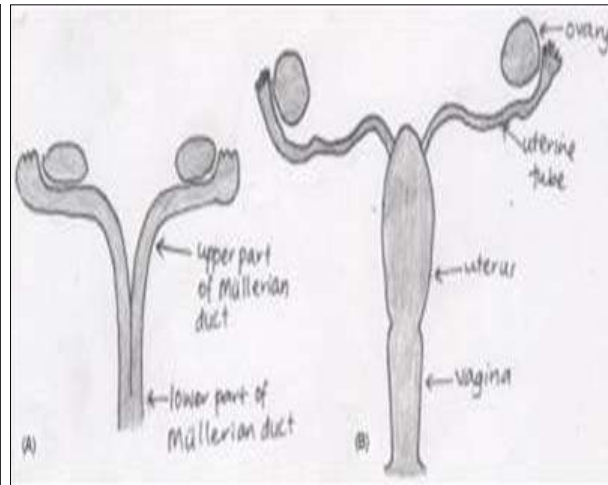
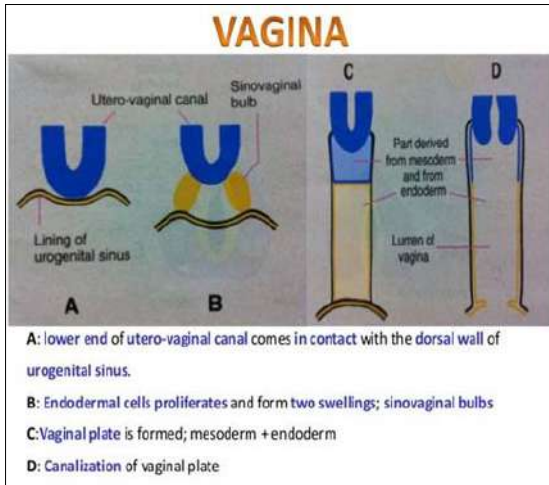


Fig. 16.18 Formation of utero-vaginal canal by fusion of the caudal parts of paramesonephric ducts.

Partially fused part: fundus of the uterus

Unfused part: uterine tube

*Thickness of myometrium increases. (Myometrium comes from the surrounding mesoderm)



Mullerian agenesis classification:

Class I: uterine agenesis/uterine hypoplasia

- a: vaginal (uterus: normal/variety of abnormal forms)
 - b: cervical
 - c: fundal
 - d: tubal
 - e: combined
- class II: unicornuate uterus/unicornis unicollis, ~15% (range 6-25%)
- a: communicating contralateral rudimentary horn contains endometrium
 - b: non-communicating contralateral rudimentary horn contains endometrium
 - c: contralateral horn has no endometrial cavity
 - d: no horn
- class III: uterus didelphys, ~7.5% (range 5-11%)
- class IV: bicornuate uterus: 2nd most common type ~25% (range 10-39%)
- a: complete division, all the way down to the external os (bicornuate bicollis)
 - b: partial division, not extending to the internal os (bicornuate unicollis)

Class V: septate uterus: commonest anomaly, ~45% (range 34-55%)

- a: complete division, all the way down to the internal or external os
- b: incomplete division, involving the endometrial cavity but not the cervix

Class VI: arcuate uterus, ~7%

Class VII: in utero diethylstilbestrol (DES) exposure (T-shaped uterus)

Diagnostic criteria

The MRKH syndrome is characterized by congenital aplasia of the uterus and the upper part (2/3) of the vagina in women showing normal development of secondary sexual characteristics and a normal 46, XX karyotype. Isolated utero-vaginal aplasia is referred to as Rokitansky sequence or to type I (isolated) MRKH syndrome. Incomplete aplasia and/or associated with other malformations, is generally referred to as MURCS association (or type II MRKH syndrome). Other associated malformations include (type II or MURCS association):

- Renal (unilateral agenesis, ectopia of kidneys or horseshoe kidney)
- Skeletal and, in particular, vertebral (Klippel-Feil

anomaly; fused vertebrae, mainly cervical; scoliosis)

- Hearing defects

- More rarely, cardiac and digital anomalies (syndactyly, polydactyly)

References

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