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RESEARCH ARTICLE

IMAGING OF RARE MULLERIAN DUCT ANOMAL (MRKH)

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ABSTRACT

Mayer- Rokitansky- Kuster- Hauser syndrome is characterized by vaginal atresia and a spectrum of uterine anomalies, including absence, hypoplasia, and duplication. Patients have a normal female karyotype, external genitalia, secondary sexual development and normal ovaries. Renal anomalies, usually agenesis and rarely ectopia or hydronephrosis, occur in about 40% of patients. Skeletal abnormality co exists in about 10% of patients. Patients with arrested development of mullerian ducts most commonly present with primary amenorrhoea.

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INTRODUCTION

It is a malformation of the female genital tract that is due to interrupted embryonic development of the paramesonephric (mullerian) ducts. It leads to hypoplasia of the uterus and the upper one-third of the vagina. It affects approximately one in 4500 live births. Ovarian function is normal; thus, patients usually present during adolescence with primary amenorrhea in the presence of normal pubertal development and secondary sexual characteristics. The aetiology is thought to be polygenic multi-factorial; occasionally, the syndrome results from a genetic mutation or deletion of genes on chromosome 16. The normal external appearance of MRKH females makes it difficult to diagnose until puberty, typically diagnosed in mid-adolescence. The average age of diagnosis is between 15 and 18 years, although occasionally a girl may be diagnosed at birth or during childhood because of other health problems. A pelvic ultrasound may be used to see the presence or absence of the uterus and its condition.

Case report

A 17 years' female presented with complaints of primary amenorrhoea. Secondary sexual characteristic was not developed. No history of abdominal pain, white discharge, burning micturition. No significant past / personal history.

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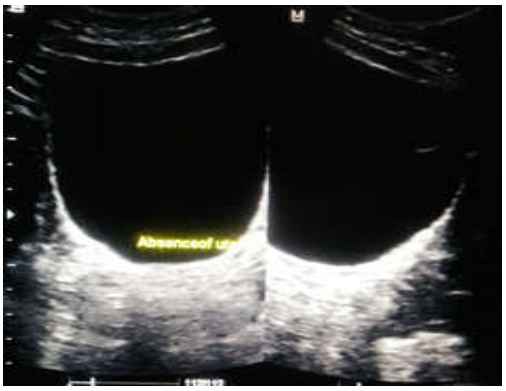
On examination: No hair growth noted in the axilla and pubic region. No breast and nipple areola complex development seen. No abnormal discharge per vagina. No hair growth noted in the axilla and pubic region. No breast and nipple areola complex development seen. No abnormal discharge P/V.

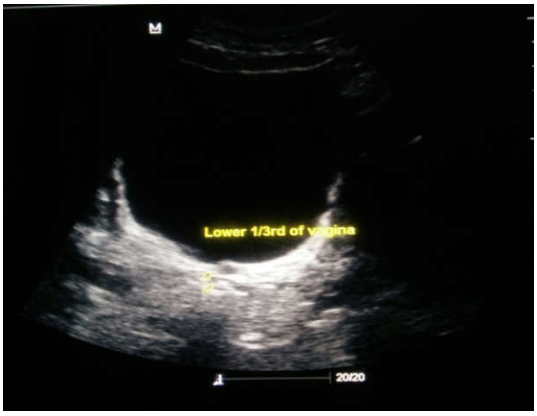
Imaging Methods

Ultrasound Abdomen and MRI Pelvis was done.

USG

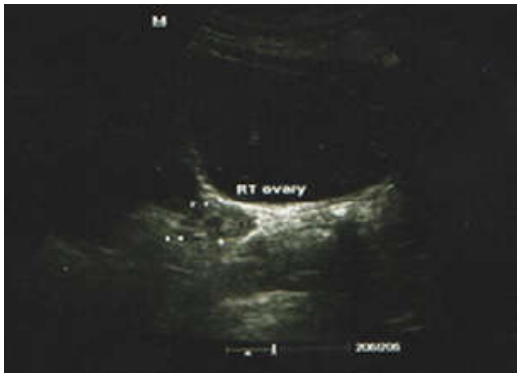
In ultrasound pelvis, lower 1/3 of vagina was visualized. Uterus was too hypoplastic to be seen in ultrasound.





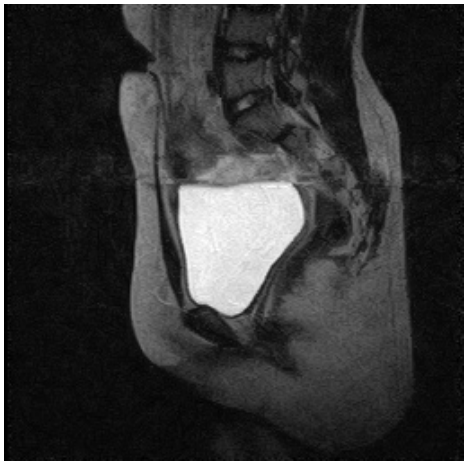
Right ovary seen and measured 1.9 x 1.5 cm.

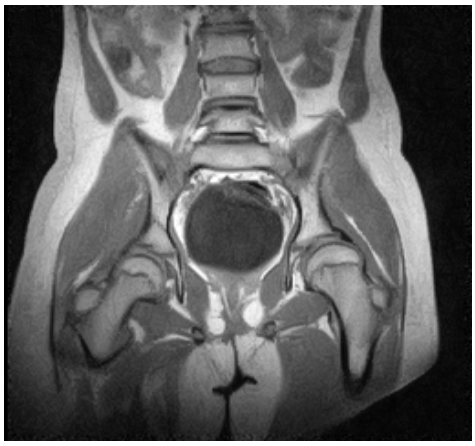
Left ovary was not visualized in ultrasound

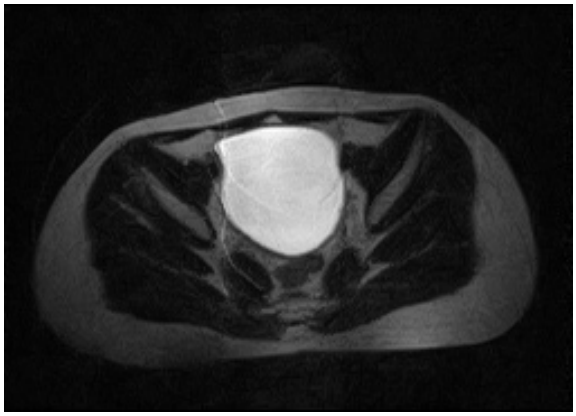


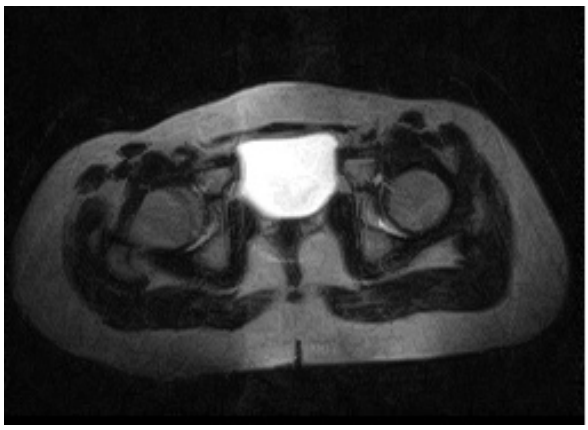
MRI

MRI Pelvis shows only lower part of uterus. Both ovaries appear normal.









DISCUSSION

Fallopian tubes, uterus, cervix and upper three fourth of vagina develops from Mullerian ducts between 8th to 12th gestational week. Developmental defect occurring at this stage leads to agenesis of Mullerian structures. The development of kidneys, ureter, and bladder occurs concomitantly at 6th-12th week. Hence renal anomalies, such as renal agenesis, ectopic kidney, fused kidney, renal hypoplasia, and horseshoe kidney are seen in 30 - 40 % of patients of Mayer Rokitansky Kuster Hauser syndrome (MRKH). Vertebral abnormalities are also found in 10 % of patients. Other rare associations are cardiac anomalies and anorectal malformations. MRKH syndrome is classified into type I and II according to accompanying urinary and other system abnormalities. Bilateral ovaries, fallopian tubes and renal systems development are normal in type I MRKH syndrome.

Complete uterine aplasia or two rudimentary horns associated with peritoneal folds are found. Lower 1/3 portion of vagina can be complete in its development, may be terminated with a blind pouch or may be atresic because it is originated from ectodermal cells. Type II MRKH syndrome may accompany with Mullerian duct aplasia, renal dysplasia and cervical sometimes anomaly with unilateral renal agenesis, renal ectopia and horseshoe kidney variation, skeletal system anomalies especially vertebral anomalies such as scoliosis, wedge shaped vertebrae, Klippel-Feil anomalies and abnormalities of extremities like syndactyly and polydactyly. MRKH has psychological consequences, but its physiological defects are surgically treatable. Surgical correction permits normal sexual function and, possibly, reproductionwith assisted techniques.

The patient may present with primary amenorrhoea and cyclic abdominal pain. Because ovarian function is normal the patient undergoes puberty with normal thelarche and adrenarche; however, menses do not begin. Patients who do not undergo evaluation for primary amenorrhea often seek clinical attention for infertility. Although the ovaries function normally, the fallopian tubes may be closed, and the uterus is often anomalous. The patient may present with inability to have intercourse. The degree of vaginal aplasia can vary from complete absence to a blind pouch. The more shallow the canal, the greater the likelihood of the patient having dyspareunia. Further investigations may reveal renal malformations like absence or ectopia of the kidneys. Some patients may present with a history of voiding difficulties or recurrent urinary tract infections and the genital anomalies may be discovered while investigating the urinary system.

Chromosomal analysis is essential to exclude karyotypic abnormalities of the X chromosome (eg, Turner syndrome). Other chromosomal aberrations may include a 46, XY karyotype, suggesting a form of androgen insensitivity syndrome (AIS). Normal circulating levels of human chorionic gonadotropin (hCG), luteinizing hormone (LH), and follicle-stimulating hormone (FSH) indicate appropriate ovarian function. Sonography is an easily accessible, noninvasive imaging modality for MRKH as it requires no radiation and is inexpensive. It easily depicts the upper level of the vagina and the length of its obstruction. It can also be used to identify uterine duplications and tubal obstruction. Simultaneously it allows assessment of the kidneys and bladder for abnormalities and visualization of some vertebral anomalies.

Magnetic resonance imaging with its multi planer capability undoubtedly provides excellent images of superficial and deep tissue planes. It can clarify "inconclusive" sonography results concerning 'cavitation' of the uterus, presence of a cervix and it improves assessment of sub peritoneal structures. The role of MRI in these patients is to depict the pelvic anatomy and to identify abnormally developed or positioned gonads. Micturting urethrography has high accuracy in demonstrating the cause, level and degree of obstruction along with features of hydro-ureteronephrosis. Conventional Pyelography can help in assessing the renal functional component besides demonstrating the pelvicalyceal system and the ureters.

Spinal radiography should be done to exclude vertebral anomalies. Laparoscopy is useful in confirmation and classification of MRKH and helps in planning the definitive reconstructive surgery.

Conclusion

The diagnosis of MRKH syndrome is usually made on the basis of clinical findings, but radiological evaluation is essential for confirmation. MRI is the imaging modality of choice. It is non-invasive and there is no use of ionizing radiation. MRI demonstration of vaginal, cervical, and uterine morphology contributes significantly to treatment planning and patient management. Sonographic study is useful to evaluate the genitourinary tract for diagnosing any associated renal anomalies.

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