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Mayer-Rokitansky-Kuster-Hauser(MRKH): Rare Disorder

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Abstract

The Mayer – Rokitansky- kuster-Hauser (MRKH) syndrome is a congenital disease in which there is aplasia of uterus and upper part of vagina in "XX" individual with normally developed secondary sexual characters. A 15 year old woman presented with pain in lower abdomen and after taking history was found to be having primary amenorrhoea. Timely evaluation and proper management of these patients is very important.

Keywords: Amenorrhoea, mullerian duct anomalies, MRKH syndrome.

Introduction

MRKH SYNDROME is a rare condition with prevalence of 1 in 4000-5000 female birth 1,2 and 2nd most common cause of defects after gonadal dysgenesis3 . It is due to incomplete development of mullerian duct .Studies are being done to find the cause but still they are unknown. MKRH usually presents as congenital aplasia of the uterus and upper part (2/3) of vagina, in young female which may include either absence or hypoplasia of uterus and fallopian tubes. The presenting complaint of the patient is amenorrhoea with normal secondary sexual characters4 . It is divided into three different types: Type1 are restricted to the reproductive system. Type2 detected by presence of asymmetric uterine remnants and abnormal uterine tubes often found to be associated with congenital renal, hearing and bone abnormalities. Type 3(MURCS type) involves utero-vaginal hypoplasia or aplasia, renal, bone and cardiac malformations1,2,3

Renal malformation includes unilateral agenesis, horse-shoe kidney, renal hypoplasia and hydronephrosis. Bone malformations include vertebral fusion, klippel-Feil syndrome and scoliosis[.1]

Case report

15year old girl presented with lower abdominal pain. History taking revealed primary amenorrhea with normal weight and height for age. The general examination was within normal range. The development of secondary sexual characters were also according to the age. Family history was not significant. In a transabdominal ultrasonography uterus was not found with difficult visualizing ovaries. MRI findings supplemented the ultrasonography observations of absence of uterus and vagina. The presence of normally appearing bilateral ovaries

showing multiple follicles was seen on MRI scan. Management in tertiary centre was planned. All other associated congenital anomalies were ruled out .The hematology profile was within normal range. Karyotyping revealed 46 XX which was normal.

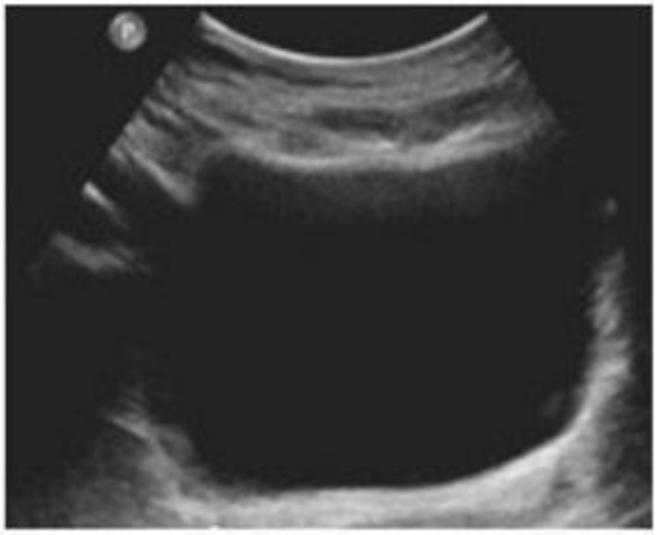

Fig. 1: USG Showing Absent Uterus.

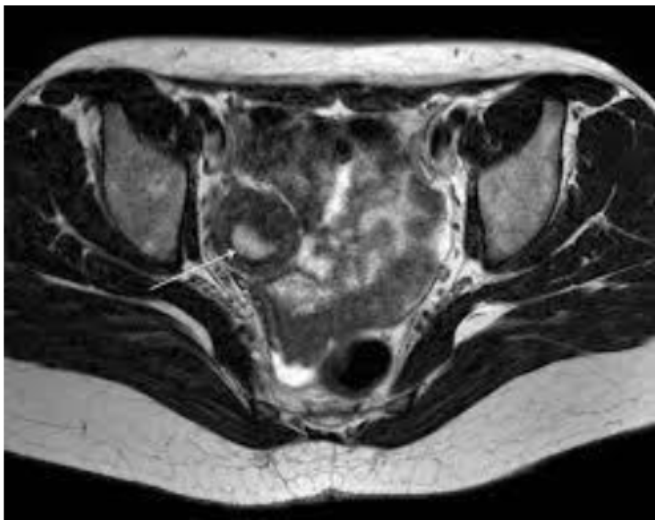

Fig. 2: MRI Image Showing Absent Uterus And Ovary

Discussion

The typical presentation of MRKH is primary amenorrhea which may be associated with colicky abdominal pain with normal secondary characters according to the age. USG is the screening method of choice which demonstrates the absence of uterus. MRI is the most sensitive and specific imaging method in the evaluation of this syndrome in both axial T1 and TSE T2 weighted scans with sagittal images to visualise uterus, ovaries and other pelvic abnormalities1,4,5,6 .Laproscopy is done after confirming MRKH by history taking examination and investigations. It causes anxiety and psychological distress in some patients so counselling of the patient is also required. Surgical creation of vagina may allow these patients to have normal sex life.

Patients who want to have children should be encouraged to adopt children take help from assisted techniques for biological offspring.

Conclusion

MRKH syndrome is one the most common causes of primary amennohea . USG is a useful for diagnosing any associated renal abnormalities. MRI is investigation of choice and precise imaging technique. Psychological stress is much more with this disorder but surgical management is good in patients for having normal sexual life, so correct evaluation of these patients and proper management is required.

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