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Case Study

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A CASE REPORT: MRKH TYPE B-MRCUS

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INTRODUCTION

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is a congenital malformation in female genital tract due to interrupted embryonic development of para-mesonephric ducts leading to uterine and proximal vagina aplasia or hypoplasia. It is of two types. Type A and Type B. Type B is atypical form and includes associated abnormalities of ovaries, fallopian tubes and renal anomalies.

RELAEVANT HISTORY

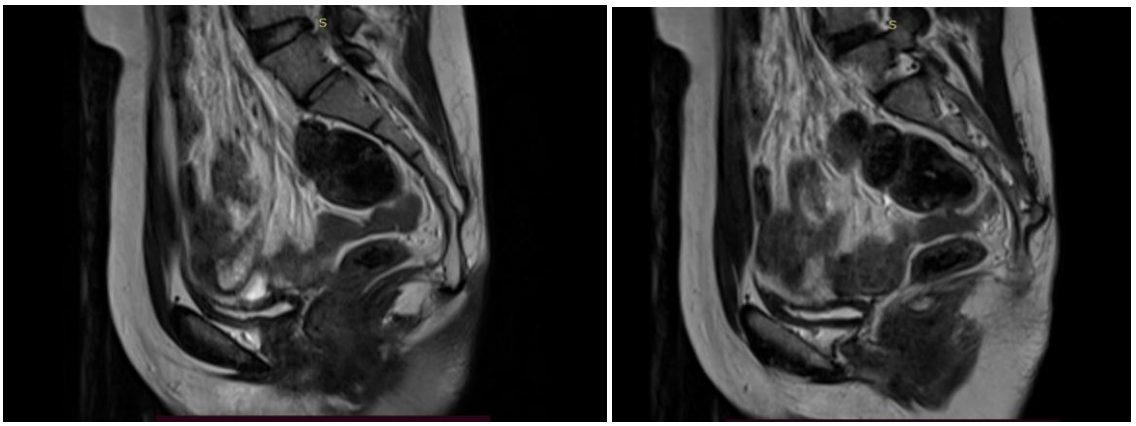
We present a case of 26/F with chief complaint of primary amenorrhea and failed marriage.

Per Abdomen: Soft and Non-tender.

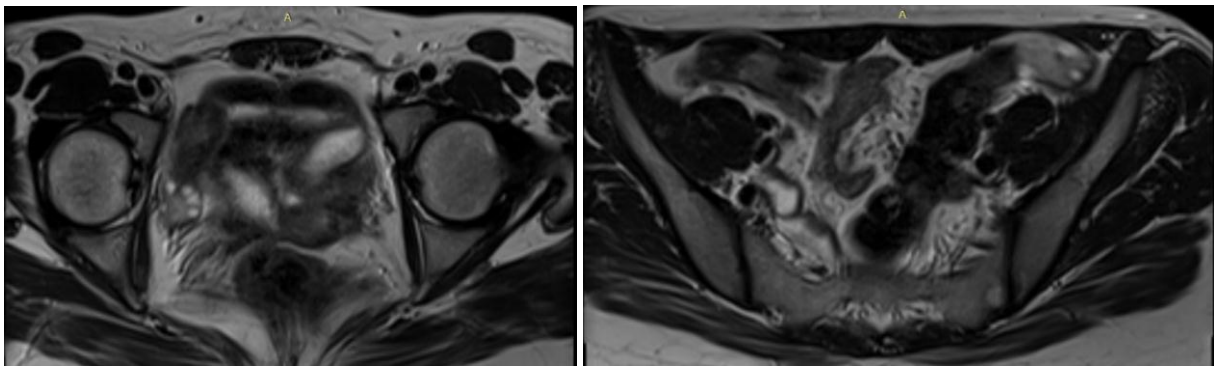
Per Speculum: Blind ending vagina.

Diagnostic laproscopy: Bilateral ovaries visualised. Uterus replaced by fibrous band and bulb like structure. Bilateral fallopian tubes not visualised. No free fluid in Pouch of Douglas.

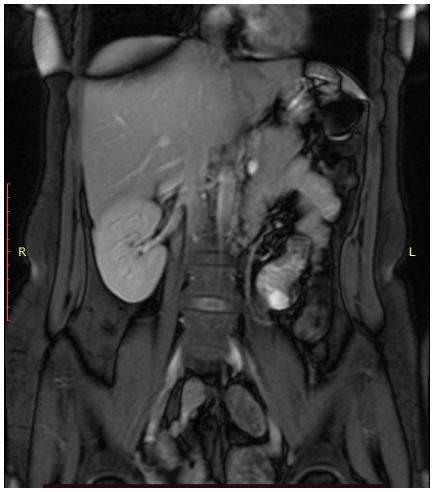
IMAGING FINDINGS

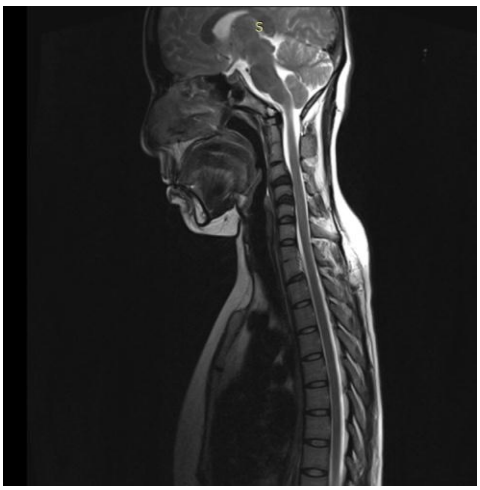


T2 SAGITTAL IMAGES: Non visualized uterus and visualized lower vagina.



T2 AXIAL IMAGES: Visualised Bilateral Ovaries.





CORONAL T2 IMAGE: Non-Visualised left kidney , SAGITTAL TE IMAGE: Fused T1and T2 vertebral bodies.

RESULTS

We were accurately able to diagnose and match the findings of diagnostic laproscopy**.**

DISCUSSION

MRKH is Class I Congenital Mullerian Anomaly. Associations of MRKH syndrome are GRES (Genital, renal and ear syndrome) and MURCS (Mullerian, Renal and Cervical Somite dysplasia)There are two forms of MRKH syndrome: a) typical form (Type-I) is characterized by only congenital absence of uterus and upper vagina with normal appearing ovaries and fallopian tubes; and b) atypical form (Type-II) includes Mullerian anomalies associated with non-gynaecological anomalies of urological, skeletal, vertebral or cardiac systems .Urological abnormalities associated with Type-II MRKH syndrome are renal ectopia, horse shoe kidney and rarely renal agenesis. Clinical presentation is characterized by primary amenorrhea and normal development of secondary sexual characteristics due to normal development of ovaries and normal ovarian functions. USG is first investigation of choice to diagnose MRKH .MRI in MRKH syndrome can not only be used as a non-invasive technique alternative to diagnostic laparoscopy and also to differentiate this entity from other possible differentials

CONCLUSION

MRKH is a congenital malformation with variable degree uterovaginal agenesis with functional ovaries. MRKH alert to look for urinary, skeletal or cardiac anomalies. MRI can diagnose MRKH and associated anomaly.

REFERENCES

1. Boruah, Deb Kumar et al. “Spectrum of MRI Appearance of Mayer-Rokitansky-Kuster-Hauser (MRKH) Syndrome in Primary Amenorrhea Patients.” *Journal of clinical and diagnostic research: JCDR*, 2017; 11(7): TC30-TC35. doi:10.7860/JCDR/2017/29016.10317.