Mayer-Rokitansky-Küster-Hauser Syndrome Complicated by Hematometra and Hematosalpinx Treated with Laparoscopic Uterine Drain Placement: A Case Report

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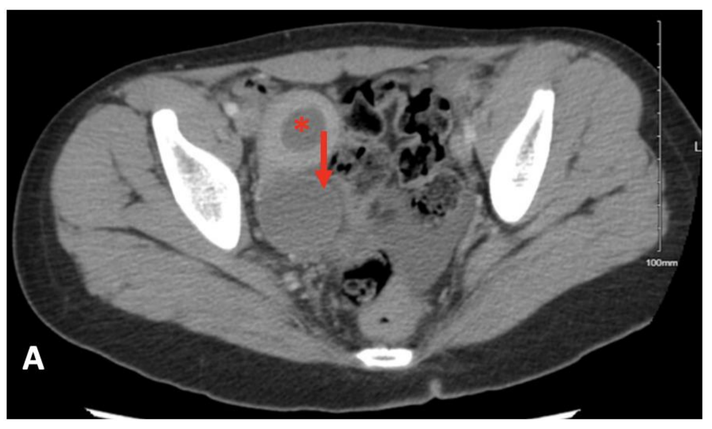
INTRODUCTION

- Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a rare reproductive tract malformation occurring in ~1:5,000 female births
- MRKH syndrome is characterized by congenital agenesis or marked hypoplasia of the Mullerian ducts derivative structures
- 3% of MRKH patients have a functioning uterus
- MRKH syndrome may be associated with skeletal and renal malformations

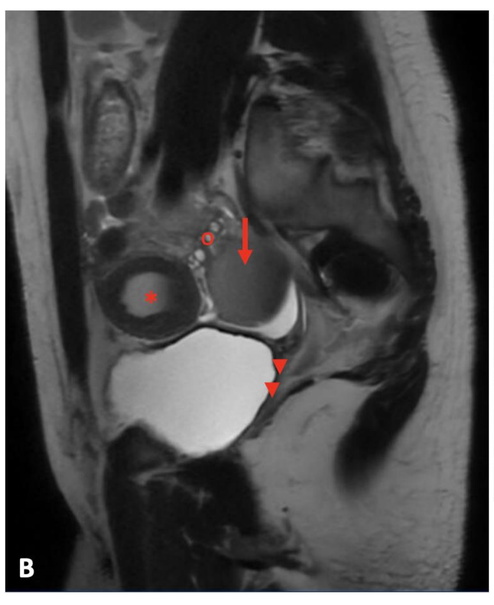
CASE PRESENTATION

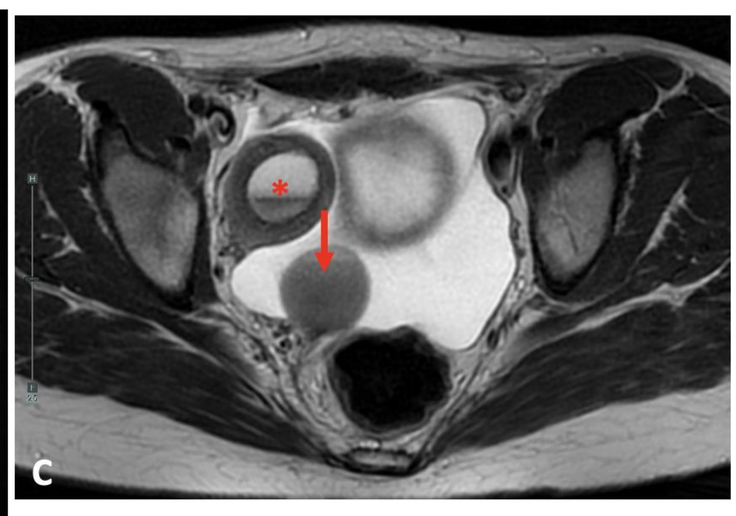
- A 12-year-old otherwise healthy female presented with persistent cyclic severe right lower quadrant pain
- She was found to have MRKH syndrome with a right-sided functioning uterus and hematometra and ipsilateral hematosalpinx
- She underwent diagnostic cystoscopy, vaginoscopy, and pelvic laparoscopy. Laparoscopic drainage of the hematometra and hematosalpinx, and hysterostomy catheter placement and uterine fixation to anterior abdominal wall were performed.
- Symptoms fully resolved post-operatively. A CT scan two weeks later demonstrated resolution of the hematometra and significant improvement in the hematosalpinx

PRE-OPERATIVE IMAGING



- (*): right uterine horn with hematometra
- (arrow): right adnexal cystic structure representing hematosalpinx
- (o): normal right ovary
- (arrowheads): cervical aplasia and vaginal atresia

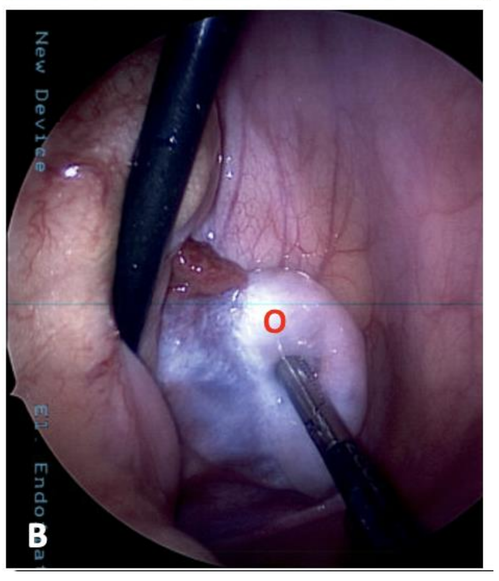




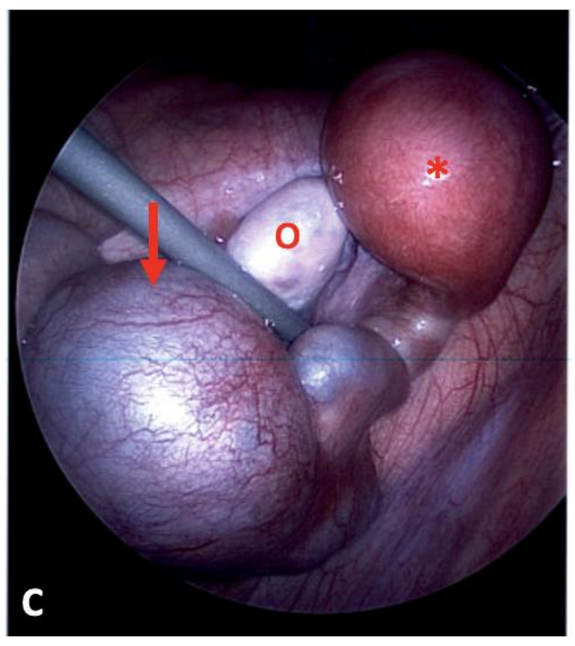
- (arrow): right supernumerary kidney

INTRA-OPERATIVE IMAGING

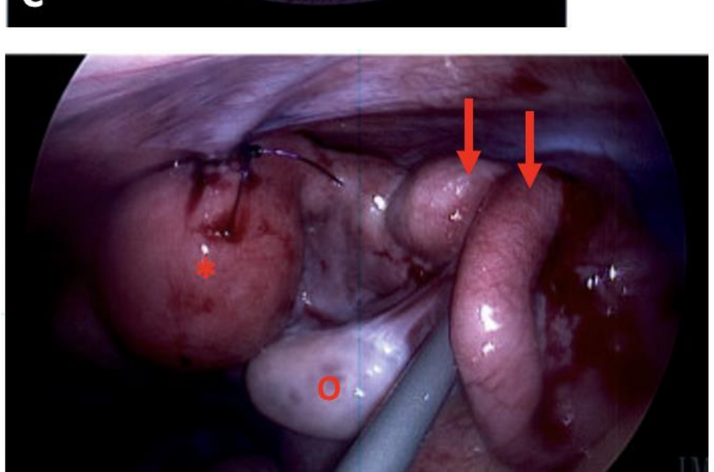
- (%): a central septum was noted, suggestive of a longitudinal vaginal septum or hemi-vaginas, was noted on vaginoscopy



- (o): left ovary located in the left lower pelvis, not attached to any tubal structure



- (*) right-sided hypertrophic uterus, significantly dilated and firm due to retained blood
- (arrow) markedly dilated and tortuous right fallopian tube
- (o): normal right ovary



- (*) uterine dome was sutured to the abdominal wall
- (arrows): right fallopian tube was decompressed after aspiration

POST-OPERATIVE IMAGING

- (*): resolution of the hematometra
- (arrow): improvement of the hematosalpinx

CONCLUSION

- In premenarchal female patients presenting with cyclic abdominal pain and amenorrhea, MRKH syndrome with a functioning uterus should be considered
- Laparoscopic uterine drain placement resulted in the resolution of clinical symptoms and radiographic findings.

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