

Obstructed Hemi-Vagina with Ipsilateral Renal Agenesis causing Pelvic Inflammatory Disease

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Background

OHVIRA syndrome is a rare mullerian anomaly with extensively varied clinical presentations making it difficult to diagnose (1). Diagnostic delays can cause pelvic inflammatory disease, sepsis and impact future fertility.

Patient Case

A fourteen year old girl presented to the emergency department with a two week history of fevers and green vaginal discharge. She had never been sexually active and had no significant medical history. She was found to be septic with a tender abdomen, malodorous vaginal discharge, elevated inflammatory markers and blood cultures positive for staphylococcus aureus.

A pelvic MRI demonstrated mullerian anomalies compatible with OHVIRA syndrome with an obstructed right vaginal cavity, multiple intra-peritoneal abscesses and a single left kidney.

The patient received appropriate intravenous antibiotic treatment and underwent a laparoscopic drainage of pelvic abscesses and incision of longitudinal partial vaginal septum. She remained well at her subsequent outpatient follow up with eventual discharge to her primary care physician at twelve months.

Diagnostic Challenges

1. Low incidence (0.1%) and lack of clinician knowledge
2. Diverse clinical presentation
3. Adolescent age of presentation causing parents/guardians and clinicians to attribute symptoms to “typical menstrual pain”

References

1. El-Gohary, M. Uterus didelphys with obstructed hemivagina and ipsilateral renal anomaly (OHVIRA syndrome: A case report. Journal of Pediatric Surgery Case reports. 2014. DOI:10.1016/j.eps.c.2014.08009
2. Bajaj S, Misra R, Thukral B, Gupta R. OHVIRA: Uterus didelphys, blind hemivagina and ipsilateral renal agenesis: Advantage MRI. Journal of Human Reproductive Sciences. 2012. DOI:10.4103/0974-1208.97811

Diagnosis & Management

Diagnosis

- MRI (2)
- Guides accurate surgical management

Management

- Based on presenting symptoms
- In case of Pelvic Inflammatory Disease – appropriate antibiotic therapy and source control through abscess drainage
- Restoration of normal pelvic anatomy through surgical management

