

Distal Vaginal Atresia

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Congenital anomalies of the genital tract are rare and present with a broad spectrum of clinical features, making their diagnosis a challenging process. Comprehensive management is imperative to preserve fertility and functionality. Therefore, correct identification is of importance for early intervention as well as keeping a high index of suspicion. Imaging can help in early diagnosis and plan the appropriate management.

A twelve-year-old female presented with multiple episodes of urinary retention associated with lower abdominal pain due to distal vaginal atresia. MRI revealed that the proximal part of the vagina was enlarged with endo-cavitary fluid collection.

The patient underwent vaginoplasty under general anesthesia. The distal portion of the vagina was reconstructed, and vaginal patency was restored successfully. The operation was followed up by manual vaginal dilation using graded dilators.

Bahrain Med Bull 2020; 42 (4): 313 - 315

The embryological development of the female genitalia is complex. Maldevelopment can result in any of the following three congenital outlet obstruction anomalies: imperforate hymen (failure of epithelial degeneration), low, mid, or high transverse septum of the vagina (incomplete unit), or atresia of the vagina¹. It is difficult to distinguish between the different types of congenital vaginal outlet obstructions, but with the advancement of radiological investigations, diagnosis of such cases has become faster and easier².

The aim of this presentation is to report a case of distal vaginal atresia and highlight the importance of early diagnosis and intervention.

THE CASE

A twelve-year-old female presented with multiple episodes of urinary retention associated with lower abdominal pain two months prior. The pain was localized to the lower abdomen and was described as dull and crampy. The patient has not reached menarche.

Clinical assessment was unremarkable as well as her past medical, surgical and gestational history.

The lower abdomen was tender upon palpation, with a palpable distended urinary bladder. No vaginal opening was seen upon examining the urogenital system.

Laboratory investigations were within normal limits. Abdominal X-ray KUB revealed a lack of pelvic gases with

faint soft tissue tumefaction in the pelvic region, see figure 1. The ultrasound revealed a large turbid fluid-filled structure posterior to the bladder, which indicates hematocolpos 5.3x2.8 cm, see figure 2. Moreover, the distal segment of the vagina was not seen. The urinary bladder, uterus, and ovarian system were not remarkable.

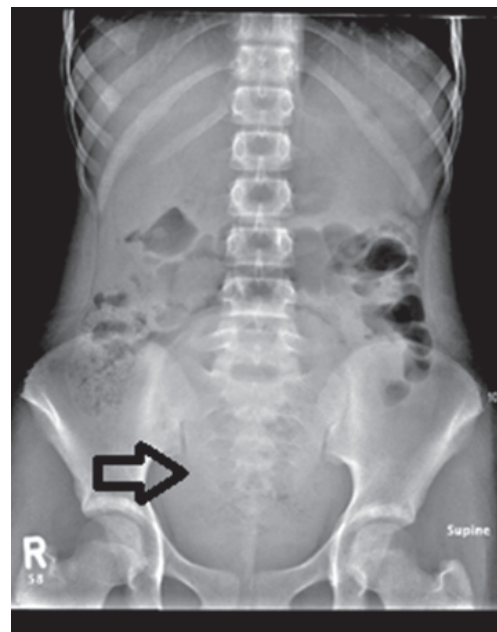


Figure 1: KUB, Lack of Pelvic Gases with Faint Soft Tissue Tumefaction

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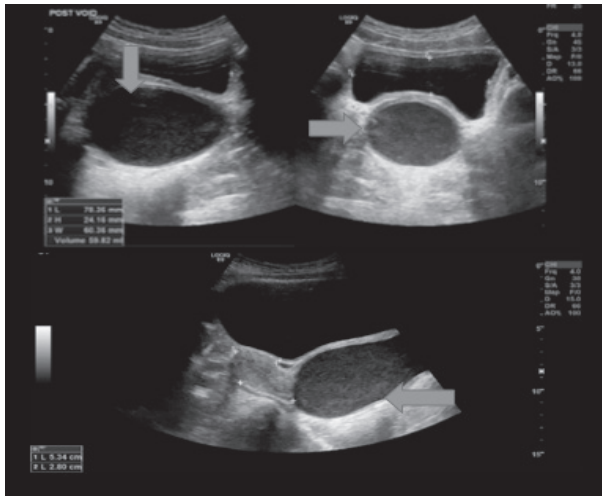


Figure 2: Pelvic Ultrasound: A Large Turbid Fluid Filled Structure Posterior to the Bladder Measuring 5.3 x 2.8 cm. Related to the Vagina Suggesting Hematocolpos

MRI revealed that the proximal part of the vagina enlarged with endo-cavitary fluid collection (T1 hyperintense, T2 hypointense-shaded) which indicated blood, and the distal end was not visualized, see figure 3-7. The uterus and cervix were displaced superiorly with the Cervix bulging into the endo-vaginal blood collection. Ovaries size and shape were normal; no ovarian pathology was identified.

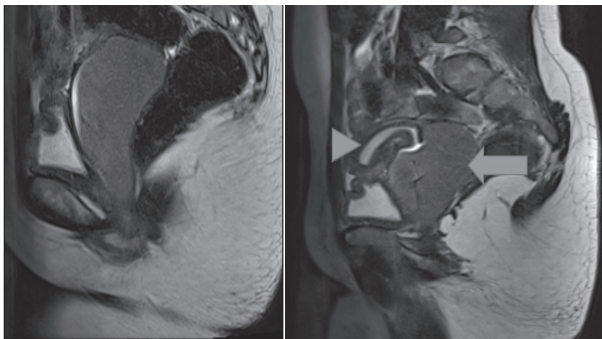


Figure 3: T2 Weighted MRI, Sagittal View Showing the Cervix Protruding into the Endo-vaginal Blood Collection (opened arrow). The Endometrium (arrowhead)

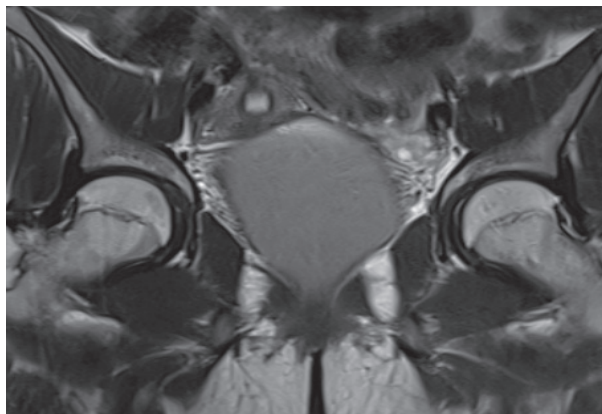


Figure 4: T2 Weighted MRI, COR

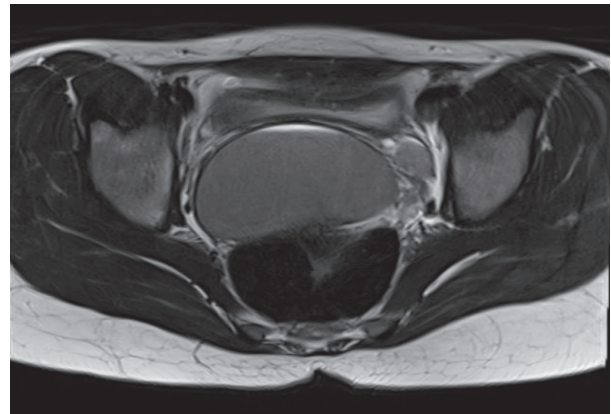


Figure 5: T2 Weighted MRI, Axial View

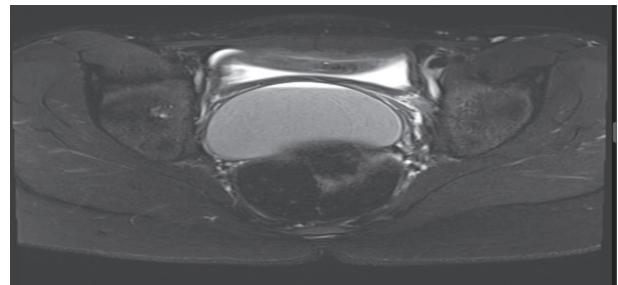


Figure 6: Fat Suppression MRI, Axial View

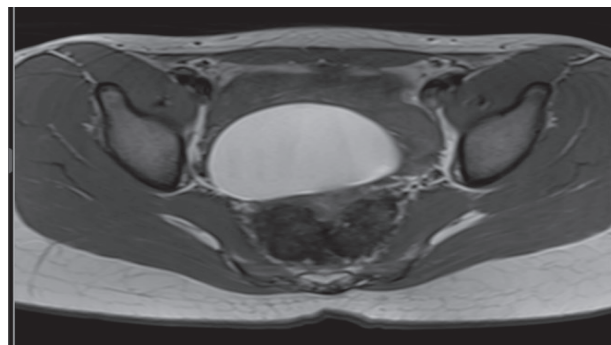


Figure 7: T1 Weighted MRI, Axial View

A diagnosis of vaginal atresia with a functioning uterus and hematocolpos was contemplated.

The patient underwent vaginoplasty under general anesthesia. The distal portion of the vagina was reconstructed, and vaginal patency was restored successfully. Postoperatively, the patient received a course of antibiotics. The operation was followed up by manual vaginal dilation using graded dilators.

On her latest appointment, the patient was doing well and tolerating vaginal dilators without any complaints.

DISCUSSION

Vaginal atresia is a congenital disorder, defined as the failure of the lower segment of the vagina to develop during the intrauterine embryological evolution of the female genitourinary tract. "Vaginal atresia is a rare condition; it occurs in 1:4000 to 1:10,000 females"⁴.

Vaginal atresia manifests during puberty, presenting with cyclical lower abdominal pain, a palpable pelvic mass and delayed menarche³. Therefore, it is crucial to keep a high index suspicion.

“The vagina originates from two embryonic structures. The upper-third from the Mullerian duct system and the lower-third from the urogenital sinus. Canalization of the vaginal canal is complete by the 20th week of gestation.”

The congenital anomalies appear when the above systems fail to fuse and canalize in a vertical plane. There are variable manifestations of Mullerian duct anomalies, vaginal atresia (VA) being one of them. In vaginal atresia, the missing portion of the vagina is replaced by fibrous tissue^{4,5}.

Early diagnosis and intervention are important to preserve the sexual and reproductive function of the patient. The spectrum of congenital urogenital anomalies is broad, and their presentation is vague and somewhat similar, which makes diagnosis very challenging. MRI takes the leading role in diagnosing and follow-up of such cases.

CONCLUSION

Genital tract anomalies are rare and variable with very similar clinical presentation. Clinicians are prompted to have a high index of suspicion when a young female patient presents with vague symptoms of abdominal pain, delayed menarche, and urinary retention. Definitive management depends on the anomaly; surgery is the main treatment modality for most cases. Postoperative follow-up is necessary until the normal menstrual cycle is established.

Author Contribution: All authors share equal effort contribution towards (1) substantial contributions to conception and design, analysis and interpretation of data; (2) drafting the article and revising it critically for important intellectual content; and (3) final approval of the manuscript version to be published. Yes.

Potential Conflicts of Interest: None.

Competing Interest: None.

Sponsorship: None.

Acceptance Date: 8 August 2020.

Ethical Approval: Approved by the Head of Scientific Development and Research, King Hamad University Hospital, Bahrain.

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