CASE REPORT

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Peritoneal Rupture of Hematocolpos in Distal Vaginal Atresia

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ABSTRACT An adolescent girl with vaginal atresia, massive hematocolpos and bilateral hydroureteronephrosis presented with an acute abdomen secondary to spontaneous rupture of the hematocolpos into the cul-de-sac. Diagnosis, treatment, postoperative course and complications of this unique case are briefly summarized. Spontaneous rupture of hematocolpos into the abdominal cavity is an extremely rare manifestation of vaginal atresia. Tertiary care management, which involves a multidisciplinary team of experienced gynecologists, plastic surgeons, urosurgeons, and critical care physicians, is recommended for optimal management of these patients. Patient education is also crucial; regular follow-ups visits and strict adherence to the postoperative vaginal dilatation schedule can reduce risk of stenosis after vaginoplasty.

Keywords: Müllerian aplasia; hematocolpos; hematometra; hydronephrosis

Vaginal atresia is a rare developmental disorder characterized by failure of canalization of the caudal portion of the sinovaginal bulb. It is associated with altered HOX gene expression and can present as a single anomaly or as a series of malformations.[1] In this article, we discuss the diagnosis, management, and postoperative course of a unique case of vaginal atresia with massive hematocolpos. The latter spontaneously ruptured into the cul-de-sac, triggering an acute abdomen.

CASE REPORT

A 14-year-old girl with vaginal atresia presented to the emergency department with acute abdominal pain and persistent diarrhea. She had a magnetic resonance imaging (MRI) two weeks ago which suggested massive hematocolpometra (20x12x10 cm) and bilateral hydroureteronephrosis (Figure 1). On examination, her blood pressure was 82/54 mmHg, her pulse rate was 130 beats per minute and her respiratory rate was 30 breaths per minute. Her abdomen was distended, tense and tender. The vaginal introitus was not present. Urethral and anal openings were closely spaced (Figure 2A). A thick pink mucous membrane covered the area of vaginal opening. An emergency ultrasound at the time was suggestive of gross hemoperitoneum. She was admitted for an emergency laparotomy.

Intraoperatively, 900 cc of old hemoperitoneum was drained. The posterior vaginal wall in the upper third was torn in two places. The uterus had reduced to a bulky size due to the outflow of accumulated blood (Figure 2B). A pull-through vaginoplasty was performed. An incision was made at the perineal end in the area of the vaginal introitus between the urethral and anal openings. Four-centimeters of the distal atretic vagina was dissected under guidance of fingers placed in the proximal vagina (inserted abdominally through the torn vagina) and rectum, respectively. The proximal vagina was mobilized and attached to the introitus. The cervix was examined at the end of the

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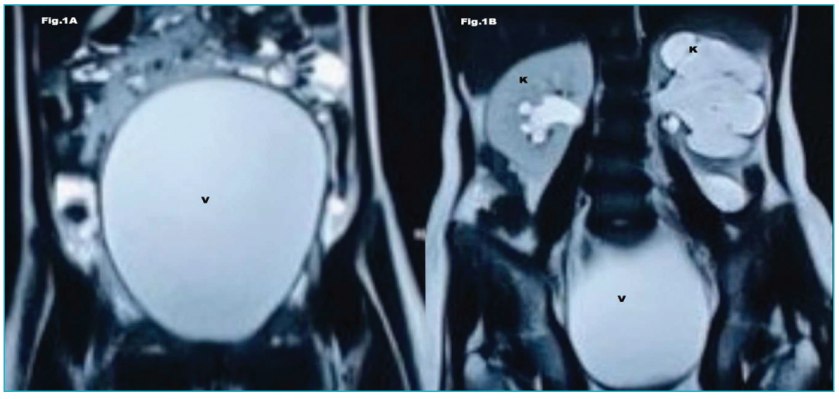

FIGURE 1: (K=Hydronephrotic kidney; V=Dilated vaginal canal) Coronal magnetic resonance imaging views showing hematocolpos **(A)** and bilateral hydronephrosis **(B).**

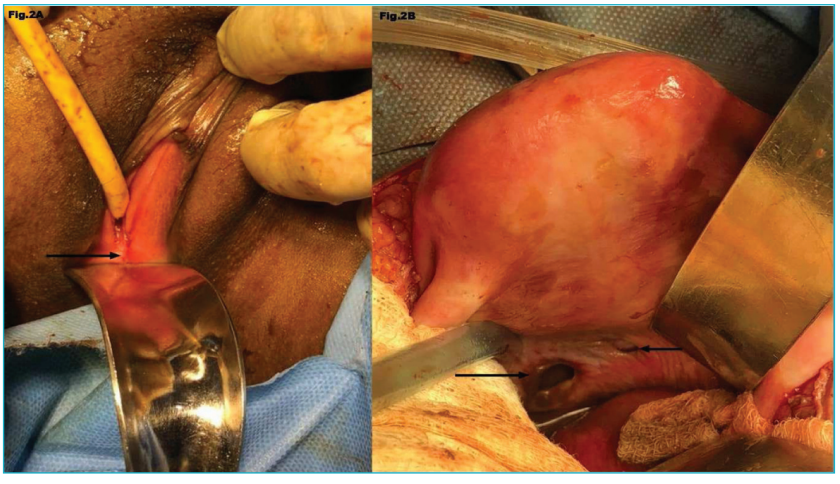

FIGURE 2: Absent vaginal introitus (arrow) **(A)**. Intraoperative finding of two posterior vaginal wall tears (arrows) draining the hematocolpometra **(B)**.

procedure and that was proper. A mold was then placed through the neovagina.

Postoperatively, she was transferred to the intensive care unit for monitoring. The exchange of vaginal molds continued weekly. She was discharged on the twenty-eighth day of surgery on gonadotropin-releasing hormone (GnRH) analogues. She was given an early consultation appointment and instructed to replace the mold herself each week. However, she was lost to follow-up during the Corona Virus Disease-2019 pandemic and presented several months later with a stenosed distal vagina. The stenosed part was resected, but this time the cervix was not visualized; therefore, the space dissected was a false vaginal passage. A second abdominoperineal reconstruction surgery was planned at a later date.

During the surgery, the abdominal cavity was opened and the base of the bladder was separated from the cervix. A 1 cm incision was made across the anterior vaginal wall just distal to the cervicovaginal junction and a Hegar #10 dilator was inserted into the vagina. Another incision was made in the area of the introitus at the perineal end and the fibrous band of tissue between the proximal and distal false vagina was resected using the dilator as a guide (Figure 3A). A split-thickness skin graft was placed over a foam vaginal mold and positioned over the raw edges of the newly formed vagina (Figure 3B). The mold was replaced after one week. On the tenth postoperative day, she

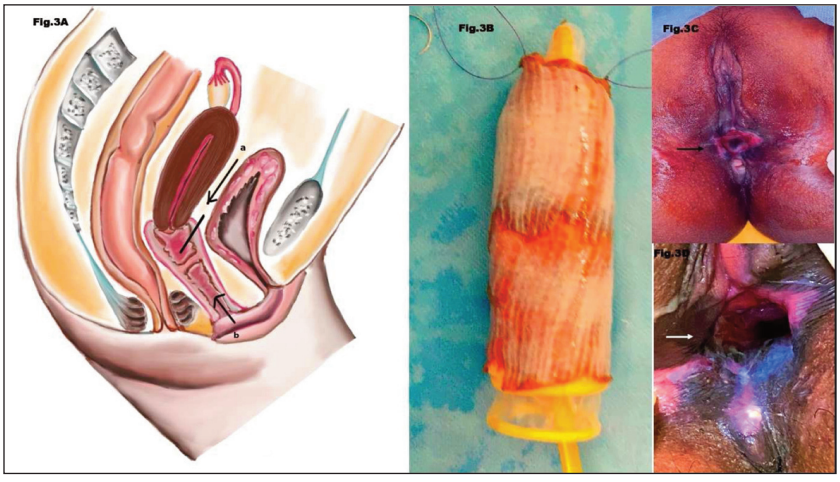

FIGURE 3: Steps of the second abdominopelvic reconstructive surgery shown as a line diagram **(A)**. After separating bladder fibers, a Hegar #10 dilator was inserted through an incision at the upper anterior vaginal wall (arrow a). At the perineal end, the fibrous band was dissected under the guidance of the dilator (arrow b). A split-thickness skin graft was placed over a foam vaginal mold that was inserted through the newly formed vagina **(B)**. The patient had a wide and patent distal vagina (arrows), after three **(C)** and six weeks **(D)** of grafting,

was discharged with a custom-made silicone vaginal mold that she could able to clean and insert weekly.

Three weeks later, graft assimilation was almost complete, covering the entire lower 5 cm of the distal vagina by 6 weeks (Figure 3C,3D). At her 6-month follow-up visit, she had regular menstrual periods and a patent cervicovaginal canal. A routine ultrasound at the time showed normal pelvic and renal architecture. She was satisfied with our treatment strategy.

The patient's guardian gave informed consent to the publication of data and to the confidentiality of her identity.

DISCUSSION

Vaginal atresia is a type I Müllerian anomaly with a reported incidence of one in 4,000 to 10,000 people.[2,3] Affected patients present at puberty with hematocolpos, which in almost all cases requires surgical drainage and vaginal reconstruction. The latter procedure is performed after careful planning in a center specializing in reconstructive surgery when the patient is mature and willing to adhere to the postoperative vaginal dilation schedule.[4-6] Until then, menstrual and pain suppression can be achieved with GnRH analogues and hormone preparations. In fact, some degree of hematocolpos is desirable during pull-through vaginoplasty as it facilitates vaginal mobilization and minimizes the likelihood of post-procedure stenosis.[6]

Contrary to the above recommendations, our patient required urgent surgical intervention as she presented with hemoperitoneum and deteriorating organ function. The hematocolpos ruptured into the cul-de-sac due to increased intravaginal pressure from the pooling of blood in the vaginal canal. Spontaneous rupture of the hematocolpos is a rare finding. In addition to Bakos et al, this is the only study documenting hemoperitoneum and acute abdomen in a patient with a congenital obstructive outflow-tract anomaly.[7,8]

The imaging modality of choice for evaluating a patient with hematocolpos is ultrasonography. However, MRI can distinguish between transverse vaginal septum, cervical agenesis, and vaginal agenesis, and rule out urogenital abnormalities.[9] Cardiac, skeletal and genetic evaluation may be performed on selected patients.

The standard surgical treatment for vaginal atresia is pull-through vaginoplasty. An introital incision is made between the area of the urethra, bladder, and rectum. Tissue planes are carefully dissected until the

obstructed proximal vagina is reached. The proximal vagina is then mobilized and placed distally at the introitus. In cases where the atretic portion is >3 cm, an interposition graft is recommended.[10] We did not use a graft in the first surgery due to anesthesia concerns. Unfortunately, this and loss of compliance predisposed her to develop vaginal re-stenosis.

Vaginal dilatation is usually recommended after vaginoplasty for abdominoperineal reconstructions, skin-flap-assisted vaginal reconstructions, and scarred vagina.[11] Literature on the exact duration, intervals, and timings of vaginal dilatation is scarce.[12] We replaced the vaginal stent with a new mold after seven days and discharged the patient. She was asked to return for mold exchanges once a week until she was sufficiently healed and pain-free. Self-dilatation was advised at the time.

A large hematocolpometra, can compress the ureters and bladder and cause hydroureteronephrosis. In advanced stages, the latter can progress to kidney failure.[13,14] This patient presented to us with severe hydroureteronephrosis. There was a transient elevation in her serum creatinine levels due to severe dehydration, which gradually improved after surgery. However, it took months for the hydroureteronephrosis to resolve.

In conclusion, peritoneal rupture of hematocolpos in vaginal atresia is a rare entity. The key to the successful management of such cases is an expert interdisciplinary team approach and comprehensive patient counseling.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Rekha Bharti, Jyotsna Suri, Zeba Khanam;* ***Design:*** *Rekha Bharti, Jyotsna Suri;* ***Control/Supervision:*** *Jyotsna Suri;* ***Data Collection and/or Processing:*** *Zeba Khanam;* ***Analysis and/or Interpretation:*** *Zeba Khanam, Rekha Bharti;* ***Literature Review:*** *Zeba Khanam;* ***Writing the Article:*** *Zeba Khanam, Rekha Bharti;* ***Critical Review:*** *Zeba Khanam, Divya Pandey, Sumitra Bachani.*

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