Case Report

A Case of Distal Vaginal Agenesis Presenting with Recurrent Urinary Tract Infection and Pyuria in a Prepubertal Girl

Ozlem Dural MD *, Funda Gungor Ugurlucan MD, Cenk Yasa MD, Ercan Bastu MD, Hulya Eren MD, Bahar Yuksel MD, Serdal Celik MD, Suleyman Engin Akhan MD

Istanbul University School of Medicine, Department of Obstetrics and Gynecology, Istanbul, Turkey

A B S T R A C T

Background: Isolated distal vaginal agenesis is a rare anomaly and mostly becomes symptomatic after menarche. We describe an unusual presentation of this anomaly in a prepubertal girl.
Case: An 11-year-old prepubertal girl presented with recurrent urinary tract infection, pyuria, and right-sided renal agenesis. The findings of perineal inspection, ultrasonography, and magnetic resonance imaging were consistent with a distal vaginal agenesis with pyometrocolpos. Discharging pyometrocolpos with dissection of the atretic portion and a pull-through vaginoplasty were performed. A cystoscopy showed no sign of a vesicovaginal or uterine fistula.
Summary and Conclusion: This rare presentation of distal vaginal agenesis reminds us that congenital malformations of the female genital tract should be considered in patients with congenital anomalies of the urinary system and/or recurrent urinary tract infection, even during the prepubertal period.
Key Words: Urinary tract infection, Distal vaginal atresia, Vaginoplasty

Introduction

Isolated distal vaginal agenesis is a rare condition characterized by normal fetal development of the uterus, cervix, and upper vagina, with absence of the lower segment of the vagina. Embryologically, the Müllerian ducts form fallopian tubes, uterus, cervix, and the proximal two-thirds of the vagina. The distal one-third of the vagina develops from the urogenital sinus. The most caudal part of the Müllerian duct start to fuse with the sinovaginal bulb at approximately 12 weeks gestational age. The vaginal canal needs to undergo a process of fusion and absorption with canalization to result in a normal vaginal cavity. Vaginal canalization is complete between the third and fifth month of embryologic development.[1] Failure of fusion or canalization of these two systems might result in distal vaginal atresia. This condition usually presents at or after menarche with cyclic and/or constant pain, a pelvic or lower abdominal mass owing to hematometrocolpos.[1] With this obstructive anomaly, hydrometrocolpos can also develop during the neonatal period and before puberty, and rarely presents with recurrent urinary tract infections, abdominal mass, and findings of obstructive uropathy. Additionally, pyometrocolpos might occur due to secondary infection of accumulated fluid.[2–4] We report a case of isolated vaginal agenesis in a prepubertal girl who presented with recurrent urinary infection and pyuria.

Case

A girl aged 11 years with a history of recurrent urinary infection and pyuria was admitted to our tertiary care center for further evaluation. The patient had three urinary tract infections in the past 6 months. *Escherichia coli* grew from a urine culture taken during this time interval. She had already received three courses of oral antibiotic therapy. She had also a history of surgery for bilateral congenital talipes equinovarus in her childhood and right-sided renal agenesis. The patient had not had her first menstrual period. On physical examination, she had Tanner stage 1 breast and pubic hair development. A perineal inspection revealed normally-developed labia majora, labia minora, clitoris, and external urethral orifice but no vaginal opening (Figure 1). The presence of pyuria was confirmed with urinary catheterization. The urinalysis result was as follows: color: dark yellow; clarity: hazy; pH: 8, glucose: negative; protein: negative; >100 white blood cells per high power field, 6-10 red blood cells per high power field. The patient was initially evaluated using transrectal ultrasonography, which showed markedly dilated upper vagina and distended uterus with echogenic debris that represented hydrometrocolpos or pyometrocolpos. Magnetic resonance imaging (MRI) was performed to delineate her pelvic genitourinary anatomy, which revealed that the endometrial cavity and upper part of her vagina was distended with fluid, which appeared hypointense on T1 and hyperintense on T2 imaging sequences (Figure 2). The length of the atretic vaginal segment was approximately 2.5 cm measured according to the MRI scan. An absent right kidney with compensatory hypertrophy of the left kidney was also observed.

The authors indicate no conflicts of interest.

* *Address correspondence to:* Ozlem Dural, MD, Sehremini Mah, Turgut Ozal Cad 34390 Fatih, Istanbul, Turkey; Phone: +905332318526
E-mail address: cinozlem@hotmail.com (O. Dural).

1083-3188/$ - see front matter
http://dx.doi.org/10.1016/j.jpag.2016.08.007

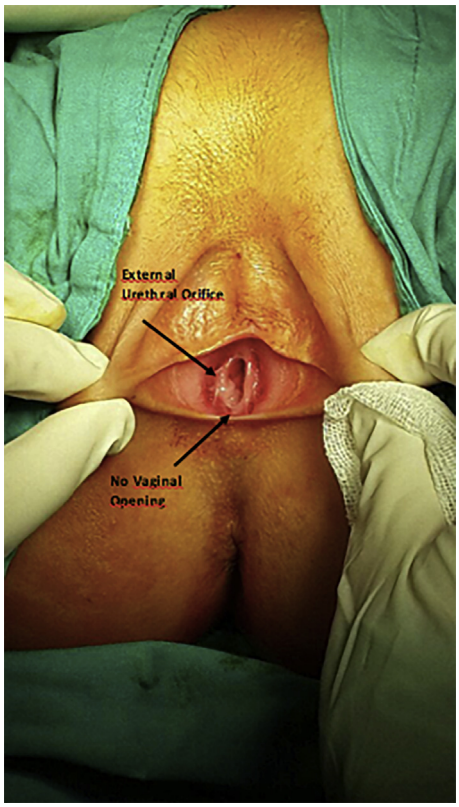

Figure 1. Appearance of the introitus.

The dissection through the atretic vaginal segment and a pull-through vaginoplasty was planned. A Foley catheter was placed in the bladder and surgical intervention was performed using ultrasound guidance. Two Allis clamps were placed at the level of the hymenal ring and a horizontal mucosal incision was performed, and careful dissection was continued between the rectum and the bladder until the bulging proximal vagina was reached. The vaginal cavity was entered sharply and the pyometrocolpos was drained and the specimen was taken from the drained pus for culture. There was sufficient proximal vagina for a pull-through vaginoplasty. The vaginal mucosa was then brought to the perineum and interrupted 2-0 Vicryl sutures were placed circumferentially. A vaginal stent coated with *nitrofurazone 0.2%* and *estriol 0.1% vaginal creams* was placed (Figure 3). After 1 week of antibiotic therapy and a sterile urine culture result, a cystoscopy was performed for suspected vesicovaginal or uterine fistula, but no abnormalities were seen. The postoperative period was uneventful and a mold was left in place all day for the first 3 months, and only at night for an additional 3 months. The patient was instructed to replace and lubricate the mold twice a day. *Estriol 0.1% vaginal cream* was applied for 3 months after the procedure. The patient was evaluated at 1, 3, and 6 months after surgery, and neither vaginal stenosis nor urinary symptoms were observed.

Summary and Conclusion

In cases of distal vaginal agenesis, the urogenital sinus fails to contribute to the upper portion of the vagina (or there is failure of canalization), and the lower section of the vagina is absent and replaced by fibrous tissue. The most common presenting symptoms are cyclic or chronic abdominal pain, and pelvic mass due to hematometrocolpos and primary amenorrhoea. This anomaly is not usually diagnosed before menarche, especially if it is not associated with other anomalies.[1]

Hydrometrocolpos might rarely occur through the accumulation of mucus during the neonatal period or just before puberty because of estrogenic stimulation of endometrial and cervical glands. Pyometrocolpos might also occur because of secondary infection of accumulated mucus. Urinary retention, obstructive uropathy, and urinary tract infection caused by compression of hydrometrocolpos or pyometrocolpos has previously been described in only a few case reports, mostly during the neonatal period.[2–4] Anomalies of the female genital tract have also been associated with other congenital anomalies such as congenital abnormalities of the urinary system, skeletal abnormalities, and imperforate anus.[5] Our patient also had a history of surgery for bilateral congenital talipes equinovarus in her childhood and right-sided renal agenesis. The presence of these anomalies, especially the existence of congenital anomalies of the kidney and urinary tract, should alert physicians to consider Müllerian anomalies.

Accurate diagnosis of underlying Müllerian anomalies is essential before any surgical intervention. Differential diagnosis of distal vaginal agenesis might consist of transverse vaginal septum at a distal level or an imperforate hymen. Genital inspection is usually beneficial in the differential diagnosis of these anomalies. Although a normal hymen with a short vagina is seen in most cases with transverse vaginal septum, in the case of distal vaginal agenesis there is no apparent vaginal opening in the perineum. However, a thick transverse vaginal septum of the lower third of the vagina can be misdiagnosed as distal vaginal agenesis. Furthermore, accurate differential diagnosis between distal vaginal agenesis and imperforate hymen is especially important to make an appropriate surgical plan. Imperforate hymen can be diagnosed by the appearance of a bluish, tense bulge at the introitus. After clinical examination, ultrasonography and MRI are mainly the preferred imaging techniques in the diagnosis of this anomaly. With MRI, the presence of a uterus, cervix, and upper vagina can be delineated, and the length of the atretic segment can be measured.[6] An MRI can also reveal detailed anatomy of the urinary system.

A variety of surgical treatments have been proposed in the literature such as pull-through vaginoplasty, using additional skin or bowel graft, and combined abdominal and perineal surgical approaches.[7] At our institution, we perform pull-through vaginoplasty with direct anastomosis of the vaginal mucosa to the mucosa at the introitus if the atretic segment of vagina measures less than 3 cm in length. For high atresias, an additional graft such as bowel or full-thickness skin graft can be used.

The timing of surgery for opening the obstruction is controversial. When the diagnosis is made during the neonatal period or early childhood, it is suggested to postpone the definitive surgery until menarche, when a