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REVIEW ARTICLE

Congenital vesicovaginal fistula: spontaneous or forced? Two cases and literature review

Hussein A. Zeineddine · Joseph Nassif · Yaser El-Hout · Fayez Suidan · Zeina M. Saleh · Tony Bazi

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Abstract
Introduction and hypothesis Congenital vesicovaginal fistula is an exceedingly rare entity. There is no consensus regarding the nature and origin of this condition. We report two cases with congenital vesicovaginal fistula and compile previously reported cases in the English literature. Theories behind the genesis of this anomaly will be briefly presented.
Methods We describe the presentation, diagnostic workup, and management of two patients with congenital vesicovaginal fistula. Previously reported cases were retrieved through an extensive English literature review using Medline and PubMed. Cases are tabulated based on the presence or absence of vaginal menstrual outflow obstruction.
Results Two women, aged 23 and 17, had had cyclic hematuria since puberty that was perceived as normal menstruation. One woman presented with an inability to have sexual intercourse, and the other with severe cyclic pelvic pain. Diagnostic workup unveiled congenital vesicovaginal fistula and distal vaginal agenesis in both. One had abnormal ureteric insertion, while the other had a history of anomalies unrelated to the urogenital system. Successful surgical correction of fistula was undertaken in both. An English literature review revealed 23 reported cases of congenital vesicovaginal fistula. While 74 % had concomitant menstrual outflow obstruction, the remaining had normal menstruation per vagina.
Conclusion Congenital vesicovaginal fistula can present as an isolated anomaly, or associated with complex malformations of a wide spectrum. The presenting symptoms as well as the age at diagnosis vary widely. While the term "congenital" implies its genesis before birth, a congenital vesicovaginal fistula can be a manifestation of faulty embryological development, but also the result of outflow obstruction.

Keywords Hematuria · Menouria · Urination disorder · Urogenital abnormalities · Vesicovaginal fistula · Vagina

H. A. Zeineddine · Z. M. Saleh
American University of Beirut Medical Center, Beirut, Lebanon

J. Nassif · F. Suidan · T. Bazi
Department of Obstetrics and Gynecology, American University of Beirut Medical Center, Beirut, Lebanon

Y. El-Hout
Department of surgery, Division of Urology, American University of Beirut Medical Center, Beirut, Lebanon

T. Bazi (✉)
American University of Beirut, 3 Dag Hammarskjold Plaza, 8th Floor, New York, NY 10017-2303, USA
e-mail: tb14@aub.edu.lb

Introduction

In 1957, Youssef first coined the term menouria to describe vesical menstruation due to a vesicouterine fistula following Cesarean section [1]. In 1971, Verhoeven [2] was the first to describe a case of primary menouria due to a vesicovaginal fistula (VVF) that he proposed to be of congenital origin. This was almost 3 decades following a report of cyclical hematuria in a woman with VVF of an unspecified etiology [3]. Surgical trauma, obstructed labor, and radiotherapy constitute the most common etiologies of VVF [4]. Congenital VVF, on the other hand, is rare [5]. The aim of this manuscript is to report 2 cases of congenital VVF and to review the English literature regarding this entity.

Case reports

Case A

A 23-year-old woman presented a few days after marriage because of an inability to have sexual intercourse. Detailed

history revealed that she had been having cyclic hematuria ever since her menarche at the age of 13 years. She had never had menstruation per vagina. She reported typical molimina every month, followed by blood in the urine for about 3 days. She also gave a history of episodes of cystitis and "kidney infections" during the first few years of puberty. She denied any history of trauma, surgery or other co-morbid illnesses. Family history was negative for the presence of similar conditions.

Clinical examination revealed well-developed secondary sexual characteristics. The pubic hair pattern was normal (Tanner stage IV) and the labia minora and majora were normal upon inspection. The introitus could not be examined owing to discomfort. The rectal examination was normal.

Magnetic resonance imaging of the pelvis revealed that the lower 4 cm of the vagina was absent. The uterus was septate, connected through a single effaced cervix to a blind vaginal pouch, measuring about 5 cm. Both ureters were asymmetrical and had abnormally low insertions just above the bladder neck. The left kidney was severely atrophic with a mildly dilated system, whereas the right kidney had multiple areas of scarring, with the right ureter distally dilated. There was evidence of multiple bilateral ovarian cysts, the largest measuring 4 cm. In order to delineate the fistulous tract, repeat MRI cuts were obtained during the "hematuria cycle." A 1- to 2-mm wide fistulous tract was visualized between the end of the vagina and the neck of the bladder (Fig. 1). Her laboratory results revealed a creatinine level of 3.2 mg/dl and blood urea nitrogen of 63 mg/dl.

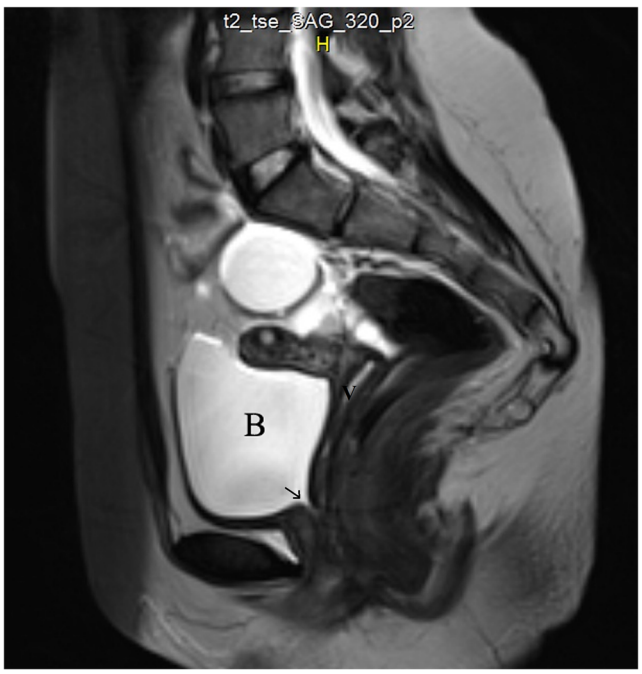

Fig. 1 T2-weighted image of sagittal MRI cut performed during "hematuria" cycle. *B* bladder, *V* proximal vagina filled with fluid of similar signal intensity. *Arrow* indicates tiny vesicovaginal fistula

After consultation with the nephrology team, the plan was surgical correction of the anomaly, consisting of the development of a functional vagina and closure of the vesicovaginal fistula, in addition to laparoscopic bilateral ovarian cystectomy. It was decided not to address the septate uterus during the same surgery, by performing hysteroscopic metroplasty through the freshly developed vagina.

Cystoscopy confirmed an abnormally low insertion of both ureters (Fig. 2). Both ostia were dilated and readily admitted the pediatric sized cystoscope. A well-epithelialized fistulous tract at the level of the bladder neck and connected to the lower end of the vagina was also seen. Laparoscopy confirmed the presence of a normal uterus, normal bilateral tubes, and bilateral ovarian cysts.

During cystoscopy, an 8-Fr Foley catheter was inserted through the fistulous tract into the blind vaginal pouch. The balloon was then inflated in order to facilitate easy localization of the blind vagina at a later stage during surgery. Vaginoplasty was undertaken by a modified McIndoe technique. A transverse H incision was made at the level of the blind hymen. Using Allis clamps anteriorly and posteriorly for traction and counter-traction, the tissue in between was sharply incised. With a finger in the rectum in order to protect from rectal injury, the urorectal space was sharply developed. This was followed by easy blunt finger dissection until the blind end of the vagina was palpated by feeling the inflated catheter balloon. Saline was injected through this catheter, filling the vaginal pouch. The latter was then incised transversely and saline was recovered. The fistulous opening on the anterior vagina was identified. A circumferential 3-mm incision was made around the Foley at the fistulous opening. Minimal dissection was performed to separate the vaginal mucosa from the vesicovaginal "fascia," and the fistula was closed in two layers using fine absorbable sutures. Next, the distal vaginal edges were easily mobilized and brought in close contact with the perineal skin, where they were circumferentially sutured in two layers. A mold was inserted into the new vaginal canal. Laparoscopic bilateral ovarian cystectomy was performed. The pathology report revealed bilateral benign serous cystadenomas. The patient had a smooth postoperative course. The Foley was removed 2 weeks later, and there was documented closure of the VVF, as no methylene blue dyed saline was recovered in the now patent vagina following bladder instillation. Since then, she has been regularly menstruating per vagina. Eight weeks postoperatively, examination revealed a well-healed perineal–vaginal suture line. An initial attempt at coitus, however, was reportedly too painful. Following many sessions of progressive perineal dilation at home, and education about conscious perineal relaxation, coitus became successful and reportedly pleasurable. In the short follow-up duration, however, no objective

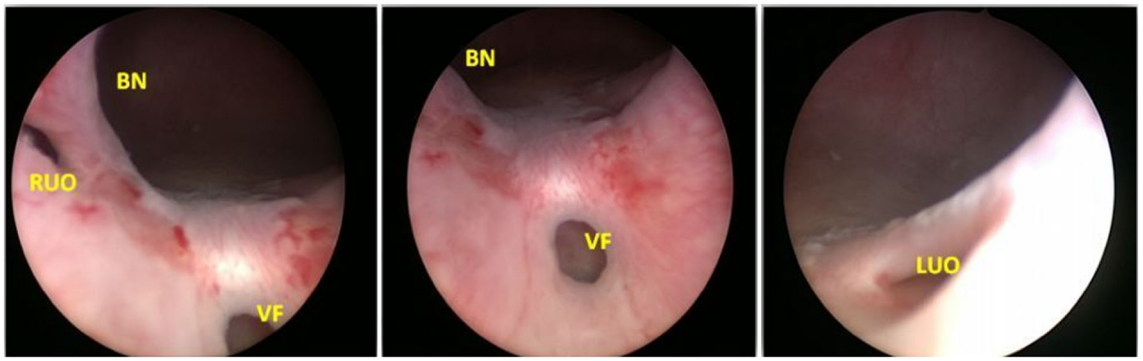

Fig. 2 Cystoscopic view of the fistula (*VF*), opening at the bladder neck (*BN*), with dilated and ectopic insertion of the right ureteral orifice (*RUO*) and of the left ureteral orifice

assessment of sexual satisfaction through validated questionnaires was performed.

Case B

A 17-year-old girl presented initially for severe cyclical pain of 3 months' duration. She reported that her "menstrual cycles" had been regular ever since menarche at the age of 12 years. On detailed questioning, however, she reported typical molimina every month, followed by hematuria, but no blood per vagina, for about 6 days. She gave history of recurrent urinary infections that responded to outpatient treatment. She also had urge and stress incontinence. Her surgical history included corrected polydactyly, cleft lip repair, appendectomy, and ovarian dermoid cystectomy.

Clinical examination revealed well-developed secondary sexual characteristics with a normal pubic hair pattern. The labia minora and majora were well developed, but the introitus was absent.

Pelvic MRI (Fig. 3) showed a significant amount of fluid in the endometrial cavity and the blind-ended upper vagina (hematometrocolpos). Both Fallopian tubes were dilated and filled with blood, the distal vagina was absent, and the ovaries were normal in size. Both kidneys and ureters were normal.

During corrective surgery, initial cystoscopy failed to reveal a fistula. The perineal area just posterior to the dilated urethral opening was incised and dissected cranially through the urorectal space until reaching the hematocolpos identified as a bulge on the anterior wall of the proximal end of the canal created. Upon pressure on this bulge, blood was emitted from the urethral meatus, suggesting the presence of a VVF. Cystoscopy was then repeated, confirming the presence of a tiny VVF. Following this, the surgery progressed as was done for patient A regarding the fistula repair and development of a functional vagina. Follow-up revealed successful closure of the VVF, and a patent vagina. During the 10-month follow-up period, she has been menstruating regularly per vagina, but has not yet engaged in coital activity. In addition, there were no recurrent symptoms of urinary infections, or urine stress and urge incontinence.

Discussion

Congenital VVF is a rare entity. It is usually associated with other urogenital abnormalities and this has been widely attributed to the close embryological relation of the Müllerian ducts, Wolffian ducts, and urogenital sinus [5]. However, the range of association with abnormalities in this entity varies extensively. Associated anomalies of the urogenital system [6, 7] as well as outside the urogenital system [8] have been reported. At the other end of the spectrum, a VVF could be an isolated anomaly [9]. Case A presents an association between a congenital VVF, distal vaginal agenesis, and a septate

Fig. 3 T1-weighted image of sagittal MRI cut revealing the uterus (*U*) and a dilated blind-end proximal vagina (*V*) filled with blood

Table 1 Congenital vesicovaginal fistula (VVF) reported in the English-language literature. Italics represent cases with no outflow obstruction

Author [reference]	Age	Clinical presentation	Associated anomalies
Whittemore [3]	21	Cyclic hematuria	Distal vaginal agenesis
			Right unicornuate uterus
			Absent left kidney
			Abdominal left 9th rib
Swinney [11]	13	Urinary incontinence	Left renal agenesis
			Left deformed uterine tube
Weiss and Dykhuizen [12]	11	Cyclic hematuria	Double uterus
			Double vagina (right vagina only is patent)
			Absent left kidney
Jones and Wheeless [13]	–	Cyclic hematuria	Distal vaginal agenesis
			Coarctation of aorta
Goodwin and Smith [14]	22	Abdominal mass	Uterine agenesis
			Left renal agenesis
			Imperforate vagina
Verhoeven [2]	21	Menouria, pyelonephritis, recurrent urinary tract infections	Hypoplasia of the right kidney
			Lumbarization of the 1st sacral vertebrae
			Vaginal atresia with hypoplasia of vaginal apex
Reed and Griscom [15]	Death at age of 3 days	–	Bilateral cystic kidneys
			Esophageal and duodenal atresia
			Imperforate anus
			Absent lower vagina
Reed and Griscom [15]	Death at age of 1 day	–	Bilateral hypoplasia
			Abnormal fascies
			Pulmonary hypoplasia
			Imperforate anus
			Absent lower vagina
Mas Torres et al. [16]	*5*	Urinary incontinence?	Left renal agenesis
Suarez and Burden [9]	*4*	Urinary incontinence	No other abnormality
Ito et al. [17]	*4*	Urinary incontinence	Left hypoplastic kidney
			Left ectopic ureteric orifice
Rousseau et al. [18]	*3*	Urinary incontinence	No other abnormality
Ishimaru et al. [19]	19	Menouria	Imperforate vagina
Asanuma et al. [20]	*4*	Urinary incontinence	Left hypoplastic kidney
			Left ectopic ureter joining with the fistula
Dolan et al. [7]	16	Urinary incontinence	Right hypoplastic kidney
			Ectopic right ureter
			Didelphys uterus
			Vaginal septum
Bandhu et al. [21]	26	Inability to achieve sexual intercourse	Partial vaginal agenesis
Chin et al. [22]	23	Menouria	Transverse vaginal septum
Srinath and Misra [23]	18	Cyclic hematuria, abdominal pain, mass in the hypogastrium	Transverse vaginal septum
Kumar et al. [24]	22	Menouria, infertility	Ectopic ureter
			Transverse vaginal septum
Rajamaheshwari et al. [8]	27	Menouria	Renal ectopia bicornuate uterus
			Transverse vaginal septum
			Skeletal abnormalities
Aseeja et al. [6]	22	Menouria	Transverse vaginal septum
Singh et al. [10]	16	Menouria	Distal vaginal agenesis
Martinez Escoriza et al. [5]	28	Menouria, impossible to undertake coitus	Distal vaginal agenesis

uterus. One may safely assume that the presence of bilateral ovarian cystadenomas is unrelated to her congenital condition. Case B, however, displays the presence of distal vaginal agenesis with congenital VVF, in addition to extra-Müllerian and Wolffian duct anomalies, namely polydactyly and cleft lip.

It is known that patients with congenital VVF may present with recurrent urinary tract infections [10]. This was noticed in both cases presented, albeit with different severities. Patient A also showed a significant decline in renal function. As suggested by the MRI imaging, there is a likelihood of chronic bilateral vesicouretral reflux as a result of abnormal ureter insertion. This reflux, along with the repetitive urinary infections, led to renal scarring and a decline in renal function. Urinary incontinence, as seen in patient B, is also known to occur with congenital VVF and can, on occasion, be the presenting symptom [7]. To further expand on the different clinical presentations and associated abnormalities, Table 1 attempts to compile all reported cases of congenital VVF in the English-language literature.

Distal vaginal agenesis is a rare abnormality of female genital malformations [25]. Following menarche, menstrual outflow obstruction leads to hematocolpos and intense cyclical pain, similar to patients with imperforate hymen. With a congenital VVF, however, the communication may allow for passage of menstrual fluid into the bladder, leading to menouria with neither hematocolpos nor intense cyclical pain [22]. In case A, the patient had been having cyclical menouria for 10 years without any intense cyclical pain. The patient did not notice any "abnormality" until her inability to consummate coitus. Patient B, however, seemed to have a rather incomplete drainage of menstrual fluid through the tiny fistula, thus leading to the formation of hematocolpos and hematometra with associated severe cyclical pain.

The Müllerian ducts (paramesonephric ducts) are responsible for the development of the uterus, Fallopian tubes and the superior part of the vagina. Endodermal outgrowths—known as sinovaginal bulbs—from the urogenital sinus are responsible for the development of the distal vagina. The patent vagina is formed upon extension and fusion of the sinovaginal bulb to the uterovaginal primordium (a tubular structure from the fusion of the paired Müllerian ducts) with subsequent canalization. Vaginal outgrowth is canalized completely at 5 months of gestation [26]. The sinovaginal bulb failed to differentiate results in distal vaginal agenesis.

It is unclear whether VVF truly is a result of an error during fetal development, or is secondary to elevated pressure of hematocolpos at menarche [5, 9, 10, 22]. In the congenital etiology scenario, one hypothesis is that during the urogenital sinus development, between the fourth and sixth weeks of embryonal life, the extreme canal of the embryonal vagina did not descend and remained in its original location, opening to the superior urogenital sinus, i.e., into the bladder [5]. Another hypothetical scenario proposes that the congenital VVF arises from duplication or early bifurcation of the ureteral bud and its incorporation into the paramesonephric duct [9].

In Table 1, we classified all reported congenital VVF cases (in the English-language literature) based on whether or not there was an accompanying outflow obstruction. Six cases (26 %) had no outflow obstruction [9, 11, 16–18, 20], whereas the remaining 17 cases (74 %) had outflow obstruction in the form of distal vaginal agenesis or a transverse vaginal septum [2, 3, 5–8, 10, 12–15, 19, 21–24]. No cases were mentioned to fulfill the criteria of a known genetic syndrome. It is noteworthy that, while such obstruction is assumed to manifest itself as the presenting symptom only after menarche, the actual genesis of the VVF could have taken place much earlier. In fact, lower genital tract secretions, resulting from exposure to maternal estrogens, do occur and may occasionally be abundant [15]. Thus, even if the patient is presenting during childhood, the fistula could have formed earlier, as a result of elevated pressure due to the secretions during fetal life and infancy. Consequently, it is entirely possible that a VVF fistula present at birth is not—per se—the result of "faulty" embryonic development, but a result of elevated pressure due to secretions induced by maternal estrogen.

Although MRI is said to be the diagnostic tool of choice regarding Müllerian fusion and mesonephric anomalies, it failed to show the fistulous tract in case B. A similar shortfall was previously reported [5]. We therefore suggest that MRI be performed during the hematuria cycle in order to provide the best valuable information in women presenting with menouria.

Vaginal reconstruction was carried out using a modified McIndoe procedure. In patients with distal vaginal agenesis and a functional uterus, the vagina should be reconstructed in a manner that would allow coital activity, and also preserve reproductive function. This may theoretically be achieved in the two patients presented. However, in case A, managing renal failure through planned renal dialysis and possible renal transplant seems to be the current priority before attempting to conceive.

In conclusion, congenital VVF is an extremely rare entity. The spectrum of associated abnormalities is wide, ranging from an isolated fistula to complex malformations. We suggest that such a fistula might be at times of congenital origin in itself and at others, a consequence of chronic outflow obstruction.

Conflicts of interest None.

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