Primary Menouria due to Congenital Urethrovaginal Fistula with Vaginal Agenesis

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ABSTRACT

Congenital Urethrovaginal fistula with vaginal agenesis is a rare variant of vaginal agenesis. It is difficult to diagnose, classify and treat because of late clinical presentation at menarche. Management is usually challenging, and it involves vaginal reconstruction.

A 22 years female presented with cyclical menouria since age of 12 years. Clinical examination revealed the absence of a vagina with developed secondary sexual characters such as axillary and pubic hair, breast development. Abdominal USG showed normal uterus and ovaries, urinary bladder with left kidney. MRI revealed functioning left kidney along with vaginal agenesis and an abnormal communication between uterus and urethra. The surgical treatment consisted in repairing the urethrovaginal fistula and vaginoplasty.

In this case, the diagnosis of congenital urethrovaginal fistula was delayed until adulthood because of vaginal agenesis. A concomitant surgery can be performed with good outcome.

Keywords: urethrovaginal fistula, vaginal agenesis, vaginoplasty.

INTRODUCTION

Congenital Urethrovaginal fistula with vaginal agenesis is a rare variant of vaginal agenesis that is difficult to diagnose, classify and treat which manifests clinically at or after menarche. We describe here a case of urethrovaginal fistula with vaginal agenesis, which has been not reported before.

CASE REPORT

A 22 years female, presented with complaints of cyclical hematuria and ammenorhea for the past nine years. Abdominal pain was cyclical and subsided with cessation of hematuria. There was no urinary or fecal incontinence. There was no history of urinary tract infection or faecaluria. Patient had undergone right sided nepherectomy at the age of three years for a nonfunctioning kidney. Physical examination revealed absence of vagina, although labia minora and majora were adequately developed. Secondary sexual growth such as axillary and pubic hair, breast development was normal. Laboratory examination revealed normal kidney function tests. Abdominal Ultrasonography showed normal uterus and ovaries, urinary bladder with left kidney. Micturating cystogram showed abnormal fistulous communication between the posterior urethra and upper vagina with contrast outlining the uterine cavity. Further evaluation revealed functioning left kidney along with vaginal agenesis and an abnormal communication between upper vagina and urethra (Figure 1).

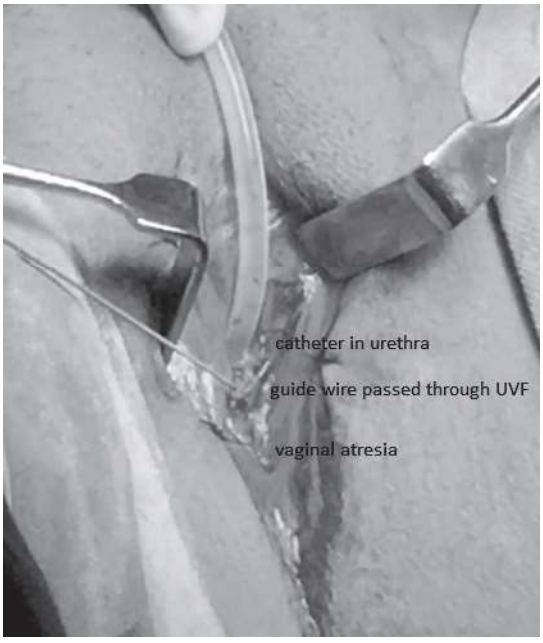

Figure 1: Showing vaginal atresia along with guide wire passed through urethrovaginal fistula.

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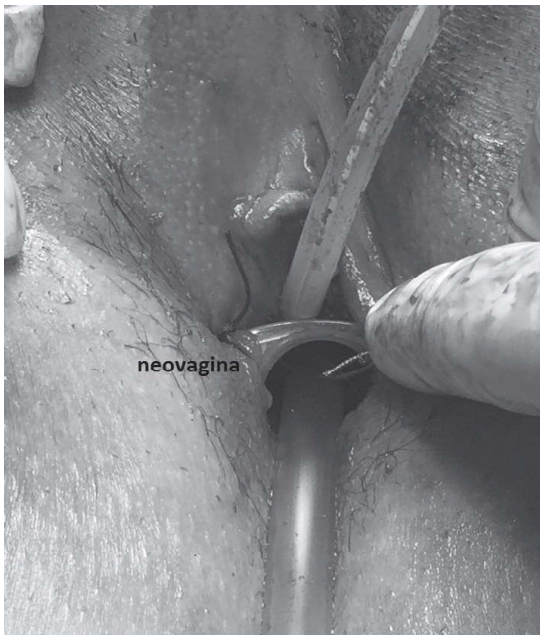

Figure 2: Showing mould passed through neovagina.

Cystoscopy performed revealed an orifice of size about 0.5 cm in posterior wall of mid urethra. Laparotomy was performed for pull through surgery for vaginoplasty from above. Intraoperative findings included normal sized ovaries, fallopian tubes and uterus. Repair of the urethrovaginal fistula with a urethroplasty was done. With a transverse incision at the level of the hymenal ring, dissection was carried out through the fibrous tissue until the upper vagina was reached, and around 50 ml of dark chocolate colored fluid was drained from uterus. The vaginal mucosa was then identified and brought down to the introitus, where it was sutured to the hymenal ring. A 5 cm vaginal mould was placed in neovagina (Figure 2), which was removed after 48 hours. The patient had received both preoperative and postoperative counseling regarding manual vaginal dilatation, future sexuality and fertility. On first follow-up, patient was taught to use vaginal dilator and advised for further follow-up. The patient is doing well as per her last follow-up at 6 months. She has started menstruating per neovagina.

COMMENTS

Congenital urethrovaginal fistula (CUVF) is very rare and is associated with embryogenic abnormality of urogenital tract.[1] In the present case, possibility could be a congenital urogenital sinus abnormality that could lead to the urethral fistula and vaginal atresia. The obstructive Müllerian anomalies true incidence is unknown, but is believed to be between 0.1% and 3.8%.[2]

The paired Müllerian ducts arise as coelomic invaginations of the mesonephros and fuse caudally to form a midline tubular structure called the uterovaginal primordium, which forms uterus and proximal vagina. Endodermal outgrowths from the urogenital sinus known as the sinovaginal bulbs give rise to the distal vagina.[3] The sinovaginal bulb extent and fuse to the uterovaginal primordium to form the vaginal plate. Subsequently, patent vagina is formed by the canalization of the vaginal plate. Failure of differentiation of the sinovaginal bulb leads to formation of distal vaginal agenesis.[4] In contrast, the etiology of CUVF remains unknown. True CUVF is of embryologic origin or results from elevated pressure of the hematocolpos at menarche remain uncleared. Oguzkurt et al[5] reported that the embryologically urethra and vagina failed to separate completely, resulting in a fistula between the urethra and vagina.

Diagnosis of CUVF can be established by cystoscopy during menouria which demonstrates the presence of the fistula.[6] MRI has emerged as a noninvasive method of diagnosing congenital UVF/VVF with vaginal agenesis.[1] The advantage of MRI is being noninvasive and gives excellent soft tissue contrast. It helps in obtaining a detailed picture of the anomaly and rule out other Müllerian fusion anomalies before any surgical intervention.

The management of vaginal atresia along with CUVF is described as closure of the CUVF and reconstruction of a neovagina.[3,4] For creation of neovagina various techniques has been described but the best option for functional outcome and sexual satisfaction with the least morbidity is still unknown. We used pull-through vaginoplasty. Using proximal vagina or peritoneum to create a neovagina is ideal because the peritoneum and the vagina are naturally moist and can provide lubrication for intercourse.[7] The thick vaginal tissue is less friable and less likely to break down during intraoperative handling and during the postoperative healing phase. It avoids the morbidity associated with the use of other grafts such as skin grafts and bowel grafts. Skin grafts (McIndoe technique) are associated with high-risk of postoperative skin scarring, contracture formation

and require long-term regular use of a vaginal mold.[8] Bowel grafts are associated with surgical risks from major bowel surgery like intestinal stenosis, intestinal wound dehiscence and fistula formation, It also has neovaginal complications of introital stenosis and mucosa prolapse. Bowel grafts also tend to have excessive mucoid discharge.[7]

With a functional uterus and distal vaginal agenesis, the aim should be to preserve the reproductive function. For our patient, pull through vaginoplasty was chosen because it would be more suitable in bridging the large gap between the upper vagina and the perineum. It avoids the problems associated with graft use from other sites and has good form and function to enable satisfactory intercourse

CONCLUSION

Congenital urethrovaginal fistulas are rare and its association with distal vaginal atresia has not been previously reported. The surgical management leads to a normal outcome.

REFERENCES

1. Singh V, Sinha RJ, Mehrotra S. Primary menouria due to a congenital vesico–vaginal fistula with distal vaginal agenesis: a rarity. Int Urogynecol J. 2011;22(8):1031-3.
2. Burgis J. Obstructive Müllerian anomalies: case report, diagnosis, and management. Am J Obstet. Gynecol. 2001;185(2):338-44.
3. Kumar S, Mandal A, Acharya N, Jain V, Kalra J, Singh S. Congenital vesicovaginal fistula with transverse vaginal septum and ectopic ureter opening in proximal vagina: case report and brief review. Int Urogynecol J. 2007;18(8):959-61.
4. Chin AI, Rutman M, Raz S. Transverse vaginal septum with congenital vesical-vaginal communication and cyclical hematuria. Urology. 2007;69(3):575-e5.
5. Oguzkurt P, Ince E, Ezer SS, Temiz A, Demir S, Hicsonmez A. Primary vaginal calculus secondary to urethrovaginal fistula with imperforate hymen in a 6- yrar-old girl. J Pediatr Surg. 2009;44:e11e13.
6. Dorairajan LN, Hemal AK. Lower urinary tract fistula: the minimally invasive approach. Curr Opin Urol. 2009;16(6):556-62.
7. Lee JM, Huang CY, Wu KY, Yen CF, Chern B, Lee CL. Novel technique of neovagina creation with uterine serosa in the treatment of vaginal agenesis associated with mullerian agenesis. Gynecol Minim Invasive Ther. 2014;3:50–3.
8. Bastu E, Akhan SE , Mutlu MF, Nehir A, Yumru H, Hocaoglu E, Ugurlucan FG. Treatment of vaginal agenesis using a modified McIndoe technique: Long-term follow-up of 23 patients and a literature review. Can J Plast Surg. 2012;20:241-4.