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Case Report

Primary cyclical menouria: a rare presentation

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ABSTRACT

Congenital vesicouterine fistula is a rare condition usually seen in association with malformations of the genitourinary tract. We report the case of a 42 yar old nulligravida with complaints of cyclical passage of blood in the urine (menouria) with severe dymenorrhoea. Physical examination revealed well developed breasts and pubic hair with a normal patent vagina but no communication between the vagina and uterus. Cystoscopy revealed the presence of a fistulous tract in the supraishtmic part between the uterus and the bladder. A hysterectomy and uterovesical fistulectomy repair were done which offered symptomatic relief to patient.

Keywords: Menouria, Vesicouterine fistula, Cervical agenesis

INTRODUCTION

The term menouria was coined in 1957 by Youssef to describe the important presenting sign of a syndrome - i.e. vesical menstruation, due to a vesico-uterine fistula and characterized by absence of urinary incontinence and absence of vaginal bleeding.[1] Vesicouterine fistula (VUF) is an abnormal communication between the bladder and the uterus. It represents a rare urogenital complication, accounting for approximately 1-4% of genitourinary fistulas.[2] Cervical atresia is an extremely rare and complex mullerian malformation.

CASE REPORT

A 42 years nulligravida married since 22 years presented with complaints of abnormal passage of blood in the urine (menouria). She had her menarche at 14 years of age which was associated with passage of menstrual blood from urethra and severe pain in the abdomen. She got married at 18 years of age. She underwent investigations at 19 years of age for primary amenorrhoea and infertility. On per speculum examination there was non visualization of cervix and ultrasonography done was suggestive of a normal sized uterus with hematometra with normal ovaries and a very small cervix non canalization. A diagnostic laparoscopy was done when she was 20 years of age suggestive of left hematosalphinx with adhesions and blood in the pouch of Douglas suggestive of endometriosis with normal uterus and normal bilateral ovaries and normal right fallopian tube. She was given injectable progesterones and did not seek further treatment for infertility. She presented to the emergency with complaints of menouria and severe dysmenorrhea. Her secondary sexual characteristics were normal with breast tanner stage 5 and pubic and axillary hair showing mature adult female pattern. On gynaecological per speculum examination there was non visualization of cervix with a normal patent vagina of length 7 cm and normal urethral opening. On bimanual per vaginal examination uterus was 6 weeks anteverted with bilateral forniceal tenderness. An contrast MRI pelvis was done which was suggestive of normal sized uterus with collection within(hematometra) with a normal vagina and bilateral hematosalphinx 9.5×5.2 cm in right adnexa and 9×6 cm in the left adnexa the fistula not being demonstrated in the study. Rest of the pelvic musculature, vasculature and organs were within normal limits. An Contrast enhanced CT urography with delayed excretory images was done on the advice of the urosurgeon, but it too failed to demonstrate the fistula. A Cystoscopy was done by the urosurgeon which demonstrated a fistulour opening near the left ureteric orifice in the vertical elevation part with a normal right

ureteric orifice. Due to severe dysmenorrhea not responding to medical management decision to do an exploratory laparotomy total abdominal hysterectomy with bilateral salphingoophorectomy with vesicouterine fistula repair was taken. Bilateral ureteric stenting with double -j stent was done prior to the procedure. A vertical incision was taken. Evidence of grade 4 endometriosis seen due to outflow tract obstruction. Total abdominal hysterectomy with bilateral salphingoophporectomy done upto the third clamp till the uterine arteries. Fistula identified just above the cervicosthmic junction dissected from bladder by urosurgeon and hysterectomy completed. Urinary bladder bivalved, fistulous tract resected, bladder sutured with vicryl 3'0 in two layers. Suprapubic catherization and foleys catherization continued for 3 weeks postoperatively. Post hysterectomy specimen histopathology report confirmed cervical agenesis.

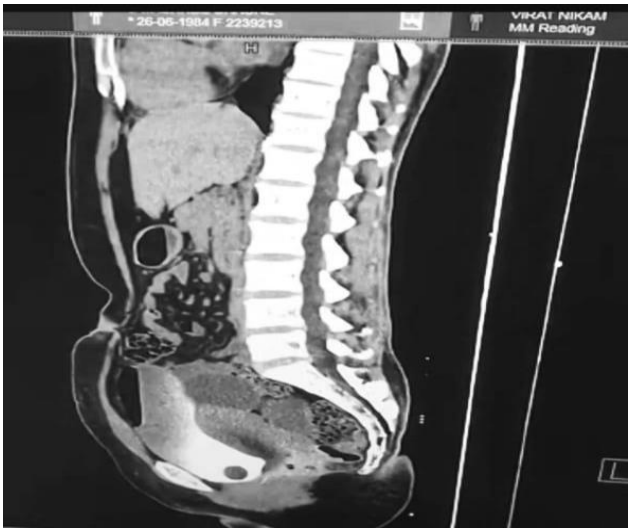

Figure 1: Sagittal section of contrast enhanced urography with delayed excretion of urograffin dye failed to demonstrate a fistula.

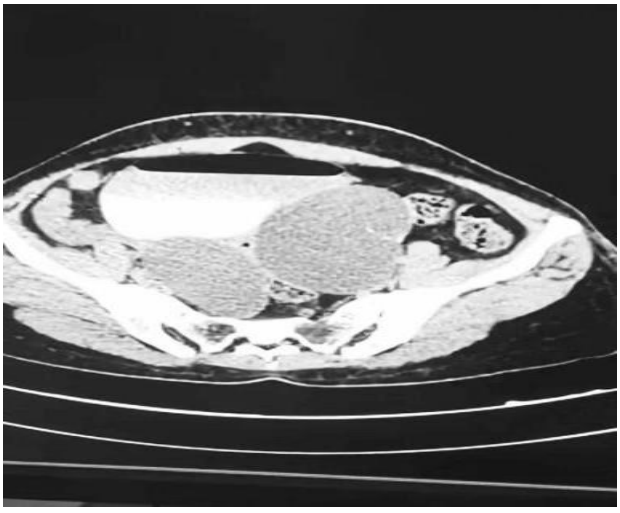

Figure 2: Axial sections of contrast enhanced CT urography showing mass of 9.5×5.2 cm in right adnexa and 9×6 cm in left adnexa.

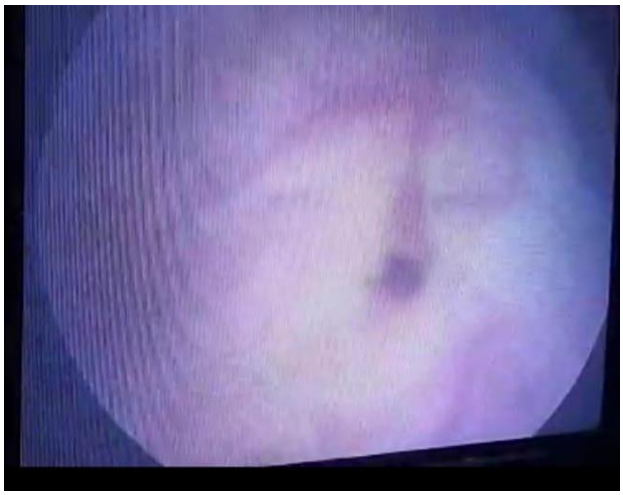

Figure 3: Cystoscopy demonstrating a fistulour opening near the left ureteric orifice in the vertical elevation part with a normal right ureteric orifice.

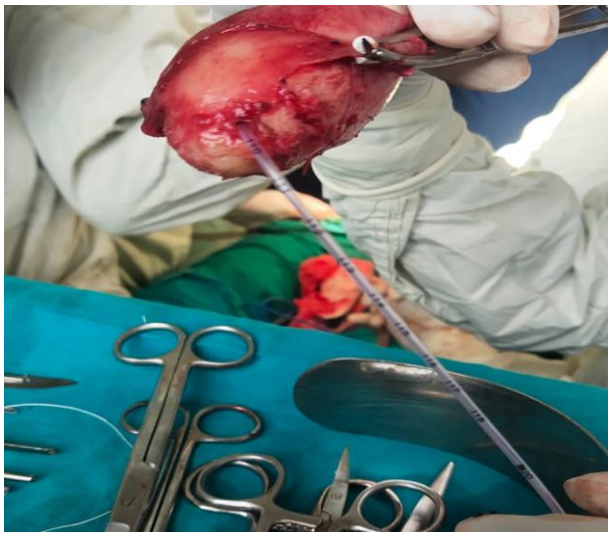

Figure 4: Post hysterectomy specimen showing cervical agenesis with infant feeding tube in fistulous tract in the supraisthmic portion.

DISCUSSION

The available literature on primary menouria is sparce. Commonest presentation of congenital vesicouterine fistula is cyclical menouria and same was seen in our case.

Presently, the main cause of vesicouterine fistula is an iatrogenic injury during cesarean section which accounts for 83-88% of cases. Other less frequent causes are inflammatory bowel disease, endometriosis, intrauterine device migration, bladder tuberculosis and congenital causes.[3,4] VUF is considered to be the least common type contrary to other types, such as vesicovaginal fistula (VVF) which is the most frequent. Congenital VVF can present as an isolated anomaly, or associated with complex malformations of a wide spectrum. Whittemore described a patient with vaginal agenesis, unicornuate uterus, a

vesicocervical communication and agenesis of the left kidney, who after surgical correction, conceived and was delivered by cesarean section.[5] Sicard described a patient with agenesis of the rectum and vagina and with a vesico-uterine communication.[6]

A classification of VUF based on the routes of menstrual flow has been proposed by Józwik and Józwik that divides VUF into three types. Type I, characterized by the triad of amenorrhea, menouria and complete continence of urine has been known as Youssef's syndrome. Type II is associated with dual menstrual flow via both the bladder and vagina. Type III is associated with normal vaginal menses and lack of menouria.[7]

Accurate and early diagnosis of vesicouterine fistula is difficult, since there are many different clinical pictures. The mainstay of diagnosis is typical history, physical examination, cystoscopy, and urinary tract imaging. MRI and contrast enhanced CT with delayed excretory images failed to make a diagnosis in our case. The available data on MRI diagnosis of VUF is very limited.[8] In a study with a small number of patients by Abou-El-Ghar et al 100% sensitivity was achieved in the diagnosis of VUF. Cystoscopy, cystography, and hysterosalpinography (HSG) play a crucial role in the diagnosis of patients with VUF. Hysterography could not be done due to cervical agenesis.[8]

Atresia or agenesis of the uterine cervix is a rare developmental malformation of the female genital tract. Fujimoto et al reported 51 cases in the world literature.[9] Patients usually present with amenorrhea and symptoms of retrograde menstruation plus hematometra. management of congenital cervical atresia is challenging. The treatment strategy should be tailored to relieve retrograde menstrual symptoms and restore fertility. Hence, a majority of clinicians view hysterectomy as the optimal primary surgical management in these patients.[10] Recently, small series have been reported which show an improved reproductive performance after utero-vaginal anastomosis.[11] The management of women with congenital cervical atresia remains controversial.

CONCLUSION

Had the patient presented early uterovaginal anastomosis with vesicouterine fistula repair could have been done was could have offered relief from symptoms and enhanced reproductive outcomes. Cervical agenesis paved the way for egress of blood through the bladder.

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