Youssef syndrome with a summary of management options

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SUMMARY

Vesicouterine fistula is one of the rare varieties of urogenital fistula. Type I urogenital fistula or Youssef syndrome is characterised by menouria, amenorrhoea and urinary continence and it mostly follows lower segment caesarean delivery. There are only scattered case reports to help guide diagnostic and therapeutic options for this condition. These patients mostly need a combination of diagnostic modalities to confirm the diagnosis. Here, we present one such case of para 4 live 4 with classical symptoms of Youssef syndrome following a laparotomy for uterine rupture repair. CT urography confirmed the diagnosis and cystoscopy helped localise the exact location. Transabdominal fistula excision and repair was done. The paper also presents a summary of diagnostic and therapeutic options for this condition as reported in previous case reports for easy reference for practising gynaecologists and urologists.

BACKGROUND

Youssef syndrome, which was a rare entity, has now become a frequent complication with increasing rates of lower segment caesarean delivery. Classically, the syndrome is characterised by menouria or efflux of menstrual blood with urine periodically, amenorrhoea and urinary continence as described by Youssef in 1957.[1] The presentation may not be typical always with some degree of urinary incontinence in few. Women might present with this complication at varied times, even years after the inciting event such as caesarean delivery. Here, we describe one such case from our institution. Also, the authors have reviewed the available literature and presented a summary of management options observed so far.

CASE PRESENTATION

A 32-year-old multiparous home maker presented to us with complaints of cyclical haematuria and amenorrhoea following a laparotomy for uterine rupture repair after vaginal birth 1 year ago. She has had two caesarean deliveries, 10 and 6 years ago, and a vaginal birth after caesarean section, 4 years ago prior to this. There was no history of any urinary leak or symptoms of urinary tract infection. General physical examination and pelvic examination revealed no abnormality and all haematological and hormonal parameters were within normal limits.

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INVESTIGATIONS

Ultrasonography (USG) of the pelvis detected no abnormality. With suspicion of urogenital fistula, she was subjected to CT urography which revealed contrast opacified fistulous tract between anterior uterine wall and posterior wall of urinary bladder measuring 1.7 cm in maximum thickness suggestive of vesicouterine fistula (figure 1).

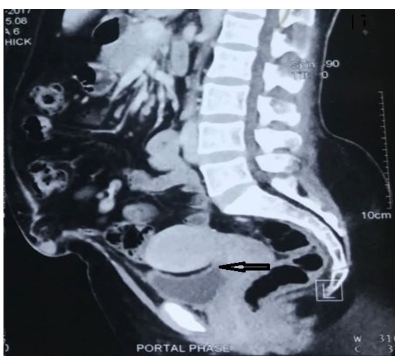

Figure 1 CT urography with contrast suggestive of vesicouterine fistula (black arrow).

DIFFERENTIAL DIAGNOSIS

The clinical impression of urogenital fistula was thus confirmed by CT urography. The authors considered vesical endometriosis as a differential diagnosis but it could not explain the reason for amenorrhoea.

TREATMENT

Management was done with a multidisciplinary approach comprising of gynaecologist and

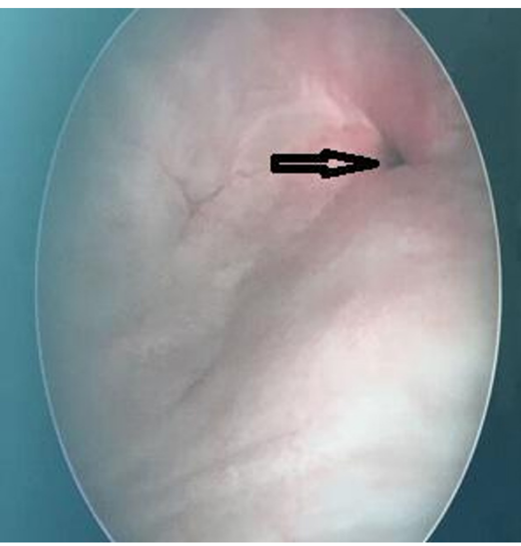

Figure 2 Cystoscopy finding of 2×2 cm supratrigonal fistula (black arrow).

Table 1 Summary of management options

Sl. no	Title	Age (years)	Symptoms	Preceding event	Diagnostic modality and finding	Management	Surgical technique	Postoperative bladder drainage	Follow-up
1	Pregnancy with neglected Youssef syndrome a case report[7]	29	18 weeks pregnancy with haematuria	Cyclical haematuria following third CS 3 years ago	MRI: linear tract visualised between the two cavities Cystoscopy: membrane seen bulging through the defect in bladder on evacuation of bladder	Laparotomy, hysterotomy and evacuation of products of conception followed by repair of uterus and bladder	O'Connor's technique with omental flap	Bladder catheter for 3 weeks	No urinary incontinence or haematuria
2	A case of vesicouterine fistula: unwanted medical anomaly but consequentiality of most-wanted medical intervention 'caesarean section'[8]	35	Cyclical haematuria, amenorrhoea and recurrent UTI since 6 years from 11th postoperative day	Second CS 6 years ago	TAS: endometrial fluid communicating with bladder IVU: right pyelonephritis Cystoscopy: fistulous opening in supratrigonal region	Laparotomy, hysterectomy and bladder repair	Trilayered closure of bladder with 2–0 vicryl	24 hours bladder irrigation with suprapubic catheter and perurethral catheter. Suprapubic catheter was removed on day 10	Cystoscopy after 2 months: healthy scar No symptoms
3	Vesicouterine fistula (Youssef syndrome): case report and literature review[3]	38	Continuous haematuria from day 1 postoperative for 3 months followed by cyclical haematuria from 6 months postoperative to 1 year. Two episodes of urethral obstruction requiring catheterisation	Third caesarean 2 years ago	Cystoscopy: fistulous opening	History of relief in symptoms for 1 year during combined oral contraceptive intake Hysterectomy and repair of bladder	Two-layered closure of bladder	Bladder catheter for 14 days	2 years since surgery: no symptoms
4	Youssef's syndrome[9]	26	Amenorrhoea and intermittent red urine since 2 ½ years came for tubal ligation reversal	Second CS 2½ years ago	Transcervical instillation of methylene blue: blue urine Sonohysterosalpingography and colour Doppler: no fluid in uterine cavity and tube but in bladder Hysteroscopy: bladder Foley's catheter seen Cystoscopy: blue dye instilled through cervix seen entering into bladder IVP: normal	Transabdominal excision of fistula and repair and tubal ligation reversal	Two-layered closure of bladder with 2–0 polyglactin and single-layer closure of uterus with no. 1 polyglactin and omental patch in between	Bladder catheter for 2 weeks	Normal menses and clear urine
5	Youssef's syndrome following cesarean section[10]	40	Urinary incontinence, haematuria and amenorrhoea	CS 1 year ago	TVS: normal Methylene blue though urethral Foley's catheter: leak though cervical os IVP: normal Cystoscopy: fistula on posterior bladder wall; catheter pushed through this came out of cervical os	Laparotomy and extraperitoneal approach for fistula excision and repair	O'Connor technique Bladder and uterine mucosa closed by two layers with 2–0 polyglycolic suture	Urethral Foley's catheter for 14 days	–
6	Post-caesarean vesicouterine fistula: Youssef's syndrome – A case report[11]	31	Cyclical haematuria and secondary amenorrhoea for 2 years	Second CS 3 years ago	USG: normal HSG: Immediate filling of bladder through fistulous tract; uterine cavity not demonstrated	Transabdominal transperitoneal repair	Bladder was repaired with vicryl 2–0 in two layers and uterus repaired in two layers	Urethral catheter for 2 weeks	Regular menstrual cycles and no urinary complaints. Postoperative HSG: normal

Continued

Table 1 Continued

Sl. no	Title	Age (years)	Symptoms	Preceding event	Diagnostic modality and finding	Management	Surgical technique	Postoperative bladder drainage	Follow-up
7	Vesicouterine Fistula (Youssef's syndrome): Imaging Findings[12]	28	Urinary leak	CS 2 months ago	Methylene blue test: positive Excretory urography: endometrial cavity opacified Pelvic CT: uterine and vaginal cavity opacification; no definite fistula Hysterography: 5 mm sized fistula between uterus and bladder Cystoscopy: fistula demonstrated	Cystoscopic fulguration of fistula	–	Transurethral catheter for 4 weeks	No urine incontinence but cyclic haematuria; open surgical repair done
8	Cystographic images of Youssef syndrome: flower on top of the bladder[13]	38	Continuous urinary incontinence for 13 years	Obstructed labour and vaginal delivery of stillborn neonate	IVU: normal upper tracts and small capacity bladder Cystogram: bilateral grade 2 vesicoureteral reflux and reflux into uterus; flower pot appearance Cystoscopy: vesicovaginal fistula and another opening into uterine cervix	Continent urinary diversion	–	–	–
9	A rare case of nocturnal urinary incontinence and menuria after lower segment cesarean section[14]	23	Chronic pelvic pain, nocturnal bedwetting and cyclic haematuria during periods since 2 years	CS 2 years ago for prolonged second stage of labour followed by bladder cathetrisation for 45 days due to haematuria	USG: normal Diagnostic hysterolaparoscopy and cystoscopy: small depression in posterior bladder wall; methylene blue injected though cervix to see spillage: negative; biopsy taken from the depression: endometrial glands Laparotomy: no fistulous tract	Injection leuprolide 1.25 mg for 3 months	–	–	After periods resumed: urinary incontinence resolved and no menouria
10	Vesicouterine fistula: Youssef's syndrome[15]	28	Cyclical haematuria (menouria) and secondary amenorrhoea since 4 years, occasional wetting of vagina	CS for non-progress of labour followed by catheterisation for 2 weeks for haematuria	USG: normal MRI: fistulous communication between bladder and uterus Cystoscopy and hysteroscopy: fistula at supratrigonal and supraisthmic region	Transabdominal tranvesical VUF repair; both ureters canalised	Uterus closed with no. 1 polyglactin suture transversely and bladder closed in two layers with no. 2 and no. 3 polyglactin sutures. Vesicouterine periotneum closed to act as transposition	Suprapubic followed by perurethral catheter removed after 3 weeks GnRH analogues given	4 weeks: MRI: well-defined hypointense scar Regular menstrual cycles and no urinary complaints
11	Treatment of vesicouterine fistula by fulguration[16]	29	Leakage of urine few days after delivery for 2 months followed by bladder catheterisation for 2 months	Second CS 4 months ago	Cystogram: fistulous tract between bladder and uterus Cystoscopy: a small epithelialised fistula orifice cannulated with 5F catheter	Fistula thoroughly fulgurated with 6F fulgurating electrode	–	Foley's catheter for 6 weeks	6 weeks: cystogram: no fistula 9 months: no urinary leakage

Continued

Table 1 Continued

Sl. no	Title	Age (years)	Symptoms	Preceding event	Diagnostic modality and finding	Management	Surgical technique	Postoperative bladder drainage	Follow-up
12	Youssef syndrome: an appraisal of hormonal treatment[17]	28 25	Case 1: amenorrhea and episodes of haematuria Case 2: amenorrhoea and cyclical menouria for 9 months	Case 1: CS followed by haematuria which cleared after a week Case 2: CS 9 months back	Case 1: IVU: leak of contrast medium into uterus during cystogram phase Hysterogram: fistula from uterus to bladder Cystoscopy: supratrigonal irregular opening of 12 mm Case 2: IVU: normal Hysterography: leakage of contrast into bladder Cystoscopy: 3 mm fistula above trigone	Case 1: levonorgesterol 0.25 mg and ethinyl estradiol 0.05 mg Case 2: levonorgesterol 0.25 mg and ethinyl estradiol 0.05 mg for 6 months	–	–	Case 1: dose doubled after 3 months due to breakthrough bleeding; did not work; surgery with transabdominal transperitoneal approach Case 2: 6 months: normal menses with non-haematuria

CS, caesarean section; GnRH, gonadotropin releasing hormone; HSG, hysterosalpingography; IVP, intravenous pyelography; IVU, intravenous urogram; TAS, transabdominal sonography; TVS, transvaginal sonography; USG, ultrasonography; UTI, urinary tract infection; VUF, vesicouterine fistula.

urologists. Intraoperatively, cystoscopy was done which revealed a 2×2 cm supratrigonal fistula 4 cm away from ureteric orifice (figure 2). A transabdominal approach was used for exploration and repair of fistula. A 2×2 cm fistula was visualised connecting anterior wall of uterus with posterior wall of bladder, which was excised. After adequate mobilisation, bladder defect was closed in two layers and uterine rent was repaired. An omental patch was placed between the two stitch lines.

OUTCOME AND FOLLOW-UP

Postoperatively, bladder catheterisation was continued for 14 days following which the patient had normal voiding habits. On follow-up, the patient was found to have regular menstrual cycles with no other complaints for 3 years now.

DISCUSSION

Since the time Youssef described a set of symptoms characterising a particular variety of vesicouterine fistula called the Youssef syndrome, many cases have been reported with similar or slightly varied features. Józwik and Józwik further analysed different types of vesicouterine fistula and classified them into three types.[2] Type I or Youssef syndrome is characterised by amenorrhoea, menouria and complete continence of urine. Type II is menouria, vaginal menses and constant or periodic incontinence of urine and type III is normal vagina menses and constant or periodic incontinence of urine. All three types more or less need similar diagnostic and therapeutic modalities. These fistulae are a result of many inciting events, most common being the lower segment caesarean delivery. It could be due to inadequate mobilisation, direct injury, suturing of bladder with uterus, devascularisation, haematoma or infection.[3] Other causes are instrumental vaginal delivery, prolonged labour, placenta percreta, anterior colporrhaphy, radiotherapy, invasive malignancy, embolisation, trauma, migration of intrauterine device, tuberculosis and cervical cerclage.[3]

The reason for urinary continence and unidirectional flow of urine and menstrual blood could be the location of fistula, that is, supraisthmic, wherein isthmus acts as a sphincter or could be due to pressure difference in the two cavities wherein the intrauterine pressure is higher than intravesical pressure.[4] When studied at microscopic level, the vesicouterine fistulae are said to have an endometrial lining at the uterine end which explains the possibility of success with hormonal management for the closure.[5 6]

The diagnosis of this condition requires high degree of suspicion as the inciting event could have happened years ago. Commonly used modalities include USG, cystoscopy, hysterosalpingography, MRI, CT urography and colour Doppler. A combination of these investigations will give a confirmation of diagnosis. Smaller fistulas can close spontaneously or with continuous bladder drainage. Other non-surgical options are hormonal treatment and cystoscopic fulguration. With surgical management transabdominal approach is required as the location of the fistula is high-up. It can be via laparotomy or minimal access. Time of repair is generally 3 months after the event to allow for the resolution of inflammation and oedema. Commonly used technique of repair is O'Connor method where fistula is excised and bladder and uterine rents are adequately mobilised and repaired individually. An omental flap can be placed between the two for better vascularisation. Postoperatively, duration of bladder drainage depends on the degree of repair and can be via per urethral catheterisation with or without suprapubic drainage.

The available literature on this condition comprise mostly of scattered case reports. The authors of this paper attempted to review about 50 such case reports. If not typical of type I vesicouterine fistula or Youssef syndrome, all cases were description of vesicouterine fistula of varying degrees. Cases representing all possible diagnostic and therapeutic options were selected and summarised for future easy reference in table 1.

Patient's perspective

I was only 32 years old and not having menses. Family, friends and some local doctors were of the opinion that I had attained an early menopause and it was okay as I already had three children. Though the recurrent blood in urine was bothering me, I was reassured that it was nothing but slight infection of the urine. Then, I came to this hospital for confirmation that it is all okay. Here, the doctors suspected some problem and got CT scan done for me. I was surprised with the diagnosis but I am happy that after the surgery I have no urinary problems with normal periods like before.

Learning points

- Vesicouterine fistula is mostly an iatrogenic complication with lower segment caesarean section as the most common cause.
- It stresses the importance of surgical skills and surgical principles.
- With type I vesicouterine fistula or Youssef syndrome the patients may not present sooner as there is no urinary incontinence.
- High degree of suspicion if required to make a diagnosis. The management is mostly surgical with excision of fistula and repair of bladder and uterus. Hysterectomy can be combined with repair if indicated.
- After histopathological evidence of oestrogen receptors in the fistula lining, hormonal management has become an area of interest.

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