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Case Report

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Surgical Management of Youssef's Syndrome: A Case Report

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Fistula · Hematuria · Amenorrhea · Continence

Abstract
Youssef's syndrome has been first described in 1957 as an atypical presentation of a vesicouterine fistula after lower segment Cesarean section. It is characterized by a triad of cyclic hematuria, amenorrhea, and absence of urinary incontinence, which is usually found in other forms of genitourinary fistulas. We describe a case report of a woman who developed a delayed Youssef's syndrome 3 months after her third Cesarean section. She was initially post-operatively treated conservatively and successfully for a bladder leakage through the wound with a bladder catheter for 1 month, and 2 months later, she developed symptoms as described above. She was successfully treated with uterus-sparing surgical fistula repair. Youssef's syndrome is a rare and atypical presentation. Physicians may easily be misled and diagnosis delayed. We describe the diagnostic approach and review the literature as to therapeutic approaches.

Introduction and Aim

Youssef's syndrome is a rare presentation of a vesicouterine fistula. It was first described by Youssef in 1957. The fistula classically presents with a triad of cyclic hematuria, amenorrhea, and absence of urinary incontinence [1]. To our knowledge, from 1974 to date, there have been only 52 cases reported in the literature. Usually, Youssef's syndrome is an iatrogenic complication following lower segment Cesarean section with an undiagnosed bladder injury.

Cesarean sections have tripled "at an alarming rate" since 1990, from 6% of all life births to currently 21% [2]. Therefore, the often not or underdiagnosed Youssef's syndrome with its atypical presentation is becoming a more common presentation, sometimes even years after the primary surgery [3].

The usual surgical treatment is resection of the bladder part of the fistula with or without hysterectomy. However, hysterectomy can be a problematic and difficult to accept choice in our cultural background. Therefore, we opted for a uterus-sparing solution. As this presentation is uncommon and atypical, we like to contribute with our case of Youssef's syndrome and present at the same time a contemporary review of the literature.

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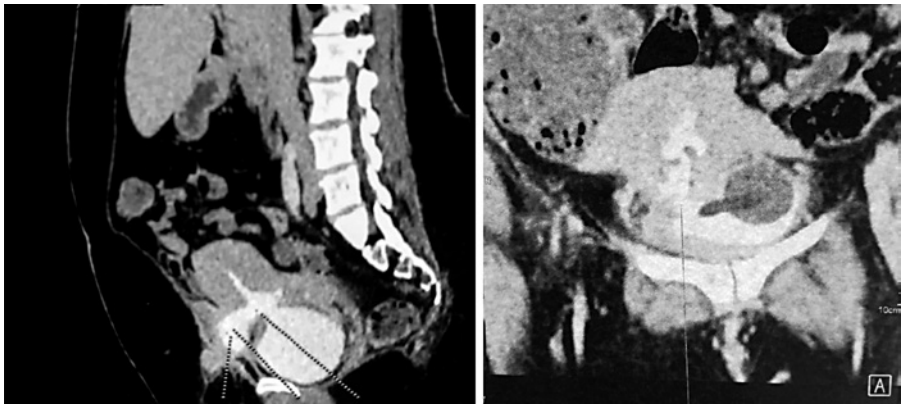
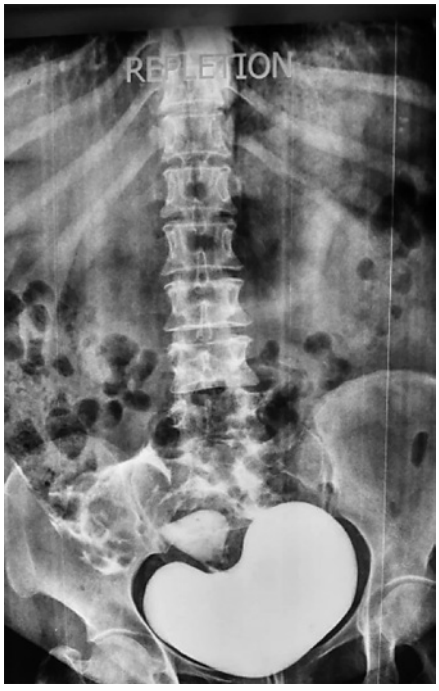


Fig. 1. Initial CT scan day 1 postoperatively, showing contrast medium extravasation from the bladder into the suprapubic space.



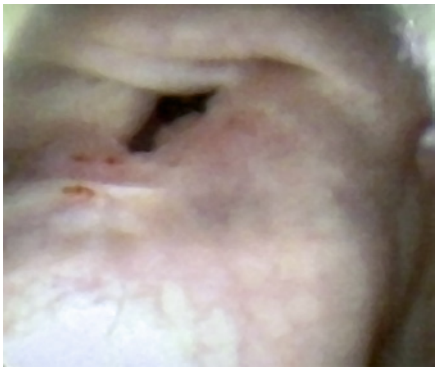


Fig. 2. Cystogram 3 months post-Cesarean section shows communication between bladder and uterus with some intraperitoneal extravasation.

Fig. 3. Cystoscopy showing a fistulous orifice at the level of the bladder fundus lateral to the right aspect of the trigone.

Methods

The patient signed a written consent for this case report. A systematic literature search was done using PubMed, Science Direct, and Cochrane databases.

Case Report

A 40-year-old woman underwent her third Cesarean section. The operation itself was documented as uncomplicated. However, on the first post-operative day, a urinary leakage started from the lower abdominal wound. A fistula between bladder and abdominal wound was suspected and confirmed by a CT scan with cystogram (Fig. 1). This was treated conservatively with a bladder catheter on continuous drainage for 1 month. After 1 month, the catheter was removed and spontaneous micturition resumed without further leakage of urine.

Three months postoperatively, the patient began complaining of cyclic hematuria and amenorrhea, with complete absence of urinary incontinence, which is the symptomatic triad suggestive of a vesicouterine fistula and Youssef's syndrome. A cystogram (Fig. 2) and a cystoscopy (Fig. 3) confirmed the diagnosis.

Because the initial conservative therapy with bladder catheter had failed and because of the delayed diagnosis with a presumably matured fistula tract by now, an indication for surgical repair was made. However, the patient wished for further pregnancies and refused hysterectomy. Therefore, a uterus-sparing approach was adopted.

Consequently, the patient underwent a laparotomy. The fistula was identified with the help of catheterization and injection of

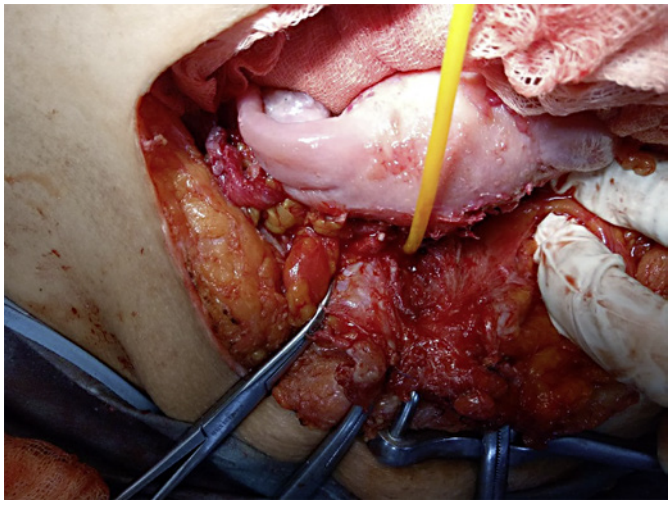


Fig. 4. The fistula has been marked with a catheter anteriorly to the uterus.

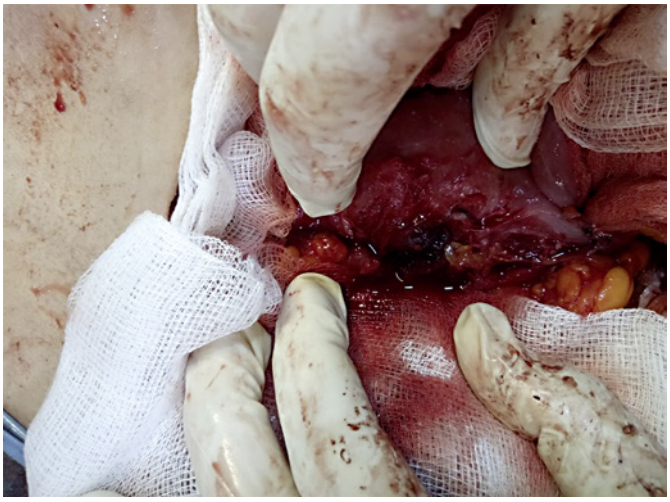


Fig. 5. The fistula has been excised in both, bladder (below) and uterus (above).

methylene blue (Fig. 4). It was excised with subsequent closure of the bladder and the uterus, which was spared (Fig. 5). An omental flap was fixed between bladder and uterus. The bladder was drained by a transurethral catheter for 15 days.

Postoperatively, the patient had no complications. She resumed her menstrual cycle after 45 days, with normal and clear micturition and full urinary continence. She remained asymptomatic without hematuria and with regular periods at 6-month follow-up.

Discussion

The first case of a vesicouterine fistula in the English literature was reported by Knipe in 1908 [4]. Cyclic hematuria was initially described by Machado [5]. Youssef in 1957 created the word "menouria" to describe cyclic hematuria and gave his name to the syndrome that incorporated the triad of cyclic hematuria, amenorrhea, and absence of urinary incontinence [1]. Although vesicouterine fistula represents only 1–4% of all urogenital fistulas, its prevalence is increasing all over the world because of the more frequent use of Cesarean section [2, 6].

Most of the vesicouterine fistulas are secondary to Cesarean sections. Several mechanisms have been proposed to explain this:

- The presence of a non-detected bladder rupture during emergency Cesarean section, usually with the fetal presentation already engaged, caused by insufficient dissection and/or insufficient drainage of the organ;
- Inadvertent application of a suture in the base of the bladder while suturing the uterus, which may also be associated with insufficient dissection/mobilization of the bladder;
- Abnormal blood supply to the base of the bladder, secondary to an abnormal vascular bed due to multiple dissections, usually after repeat Cesarean sections [7].

The atypical symptomatic triad of Youssef's syndrome is explained by an intact cervical-isthmic uterine sphincter. When the tone of the intact sphincter has recovered after Cesarean section, the menstrual blood during menses then enters the bladder following the way of least resistance.

The patient then presents with cyclic hematuria ("menouria"), amenorrhea (blood exiting through bladder), and absence of urinary incontinence (since both sphincters in cervix and bladder remain intact) [8]. The latter is otherwise typical for other forms of urogenital fistulas, and it is this lack of incontinence which most often delays the correct diagnosis of a fistula.

Once suspected, the diagnosis can be confirmed by cystography, intravenous pyelography, hysterography, sonography, and other types of imagining exams (i.e., CT scan with bladder filling) [9]. Cystoscopy will identify a fistulous opening in most cases or erythema and pneumaturia.

We reviewed 31 cases of Youssef's syndrome that were reported in the literature between 1985 and 2018. In two-thirds of patients (68%), a uterus-sparing repair was per-

formed by laparotomy. In 4 of those women, a complete bladder dissection was needed to reach the fistula.

In 16%, hysterectomy was performed. In 3%, a transvaginal surgical approach was used. In each 6% of women, hormone therapy was administered or they refused any treatment [3, 5–9]. Evidence is emerging that hormone treatment may be a promising alternative in selected cases. This consists of a 6-month amenorrhea induced by administration of luteinizing hormone-releasing hormone analogue [10].

As the worldwide rate of Cesarean sections continues to rise, the incidence of Youssef's syndrome will increase as well. It is important that surgeons are aware of the atypical triad of cyclic hematuria, amenorrhea, and absence of urinary incontinence associated with Youssef's syndrome. This will allow appropriate diagnostic and curative procedures to be performed in a timely manner. Surgical management remains the gold standard for treatment of Youssef's syndrome and has a good prognosis.

Statement of Ethics

The patient signed a written consent for this case report and agrees to the publication of details and photos related to the case.

Conflict of Interest Statement

The authors have no conflicts of interest to disclose.

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The authors did not receive any funding.

Author Contributions

R.K. and A.M. collected the case data and wrote the first draft. N.N.P.B. edited and submitted the final draft.

References

1 Youssef AF. Menouria following lower segment cesarian section: a syndrome. Am J Obstet Gynecol. 1957;73:759–64.
2 Optimizing Caesarean section use. [cited 2020 Mar 27]. Published October 12, 2018. www.thelancet.com/series/caesarean-section.
3 Tancer ML. Vesicouterine fistula: a review. Obst Gynecol Surv. 1986;41(12):743–53.
4 Knipe WHW. Vesico-uterine fistula. Am J Obstet. 1908;57:211–217.
5 Machado L. Fistulas vesico-cervicais apos cesareana segmentar. Rev Gynec Obstet. 1935;9:471.
6 Porcaro AB, Zicari M, Antoniolli SZ, Pianon R, Monaco C, Migliorini L, et al. Vesicouterine fistulas following cesarean section. Report on a case, review and update of the literature. Int Urol Nephrol. 2002;34:335–44.
7 Machado RA Jr, Machado LC Jr, Lourenço LL. Vesicouterine fistula (Youssef syndrome): case report and literature review. Rev Bras Ginecol Obstet. 2018;40(9):563–9.
8 El Fassi MJ, Tazi K, Karmouni T, Koutani A, Ibn Attya AA, Hachimi M, et al. Youssef's syndrome (vesico-uterine fistula): about three cases. Ann Urol. 2003;37:184–6.
9 Rao MP, Dwivedi US, Datta B, Vyas N, Nandy PR, Trivedi S, et al. Post caesarean vesicouterine fistulae: Youssef syndrome: our experience and review of published work. ANZ J Surg. 2006;76(4):243–5.
10 Yokoyama M, Arisawa C, Ando M. Successful management of vesicouterine fistula by luteinizing hormone-releasing hormone analog. Int J Urol. 2006;13(4):457–9.