Congenital agenesis of seminal vesicle

Hong-Fei Wu, Di Qiao, Li-Xin Qian, Ning-Hong Song, Ning-Han Feng, Li-Xin Hua, Wei Zhang

Department of Urology, First Affiliated Hospital of Nanjing Medical University, Nanjing 210029, China

Abstract

Congenital agenesis of the seminal vesicle (CASV) is frequently associated with congenital absence of the vas deferens (CAVD) or ipsilateral congenital vasoureteral communication. We reported two cases of a rare condition that the vas deferens open ectopically into Mullerian duct cyst associated with agenesis of the ipsilateral seminal vesicle. The diagnosis was confirmed by vasography. Transurethral unroofing of the Mullerian duct cyst was performed in both patients with favourable results, however, assisted reproductive technology (ART) was still necessary for them to father children. (Asian J Androl 2005 Dec; 7: 449–452)

Keywords: seminal vesicle; vas deferens; congenital abnormalities; vasography

1 Introduction

The seminal vesicles can be congenitally absent at one or both sides [1]. The incidence of unilateral agenesis of the seminal vesicle is 0.6 %–1 % [2]; while the incidence of bilateral agenesis is unclear. It is known that congenital agenesis of the seminal vesicle (CASV) may occur in patients with congenital absence of the vas deferens (CAVD) or congenital vasoureteral communication [1]. CAVD is frequently found in association with mutation(s) of the cystic fibrosis transmembrane regulator (CFTR) gene [2–6]. While ectopia of the vas into ureter is caused by incomplete absorption of the common Wolffian duct [7], it is safe to say that CASV is associated with CFTR gene mutations and defects in the mesonephric duct. Herein we presented two cases of CASV associated with ipsilateral ectopic vas into the Mullerian duct cyst, which may complement what are already known on CASV.

2 Case reports

2.1 Case 1

A 37-year-old man presented for infertility. Physical examination revealed the right vas deferens was absent with only a 5-cm-long rudiment ended in a node approximate to the epididymis cauda, and the whole epididymis was enlarged. The right testis and the left scrotal contents were normal. Digital rectal examination was unremarkable. Serum testosterone (T), follicle-stimulating hormone (FSH) and luteinizing hormone (LH) were normal. Semen analysis yielded azoospermia with low seminal volume of 1.2 mL, low pH of 6.5 and a few erythrocytes. Semen fructose test was negative. Ultrasonography and computed tomography (CT) demonstrated that the kidney, ureter, and bladder were normal, except for a cyst inside the prostate on the midline (Figure 1 A), with a diameter of about 1.0 cm; both seminal...
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vesicles were absent. Bilateral testicular biopsy revealed normal spermatogenesis. Left vasography showed the vas deferens terminated ectopically into a cyst located at the prostatic midline, as well as an absent left seminal vesicle (Figure 1 B). Regarding the midline location of the cyst, we considered it as Mullerian origin with respect to the X-ray appearances of Mullerian duct cyst proposed by Hendry and Pryor [8]. Transurethral unroofing of the Mullerian duct cyst was performed. On histological examination the cyst wall was lined with low hyperplastic columnar and cuboidal epithelial cells. Semen analysis 2 months after the operation demonstrated asthenospermia (sperm motility of 20 %, and very poor forward progression) with seminal volume of 2.0 mL, pH of 6.4 and sperm density of 45.48 × 10^6/mL, with no erythrocytes could be found. Semen fructose test was still negative.

2.2 Case 2

A 28-year-old man complained of hematospermia and infertility. Physical examination was unremarkable. Serum T, FSH and LH were normal. Semen analysis yielded azoospermia and hematospermia. Semen fructose test was equivocal. Ultrasonography and CT demonstrated the kidney, ureter and bladder were normal, except for a cyst inside the prostate on the midline (Figure 2 A), with a diameter of about 1.5 cm. The right seminal vesicle was absent and the left, hypotrophic. Bilateral vasography revealed an ectopic right vas deferens ended in a Mullerian duct cyst associated with the absence of ipsilateral seminal vesicle, the left vas deferens crossed over the midline, emerged with a hypotrophic seminal vesicle and the ejaculatory duct terminated into the Mullerian duct cyst (Figure 2 B). Transurethral unroofing of the Mullerian duct cyst was performed and histological examination was consistent with the diagnosis. Antibiotic therapy was applied pre- and postoperatively. One month later, the symptom of hematospermia disappeared. Semen analysis 6 months after the operation revealed asthenospermia (complete absence of sperm motility) with seminal volume of 1.6 mL, pH of 7.4 and sperm density of 36.73 × 10^6/mL. Semen fructose test was mildly positive.

3 Discussion

Anomalies of the seminal vesicles can be categorized into abnormalities of number (agénesis, fusion, duplication), maturation (hypoplastic), position (ectopia) and structure (diverticulum, cyst, communication with the ureter) [1]. Their significances lie in their frequent associations with mal-development of other mesonephric derivatives, such as the vas deferens, ureter and kidney. Among these abnormalities, agénesis of the seminal vesicle is perhaps the commonest. It was widely accepted that if an embryological insult occurs before 7 weeks’ gestation when the ureteral bud separates from the mesonephric duct, the seminal vesicle anomaly may be associated with renal malformation. If the insult occurs after 7 weeks’ gestation, the seminal vesicle agénesis will not be associated with renal agénesis [2].
The etiology of CASV was not known, but it was often found in patients with CAVD [2–6] or ipsilateral ectopia of the vas deferens [1, 7, 9–13]. Therefore, defects in the mesonephric duct and mutations of the CFTR gene are probably the main causes of CASV. Although it was said that CASV might occur in patients with renal agenesis [1], no cases of CASV have been found with normal developed vasa deferentia.

Previous studies suggested that in patients with congenital bilateral absence of the vas deferens (CBAVD), the incidence of bilateral CASV was 23 %–43 % [3–6, 14] and the incidence of unilateral CASV was 27 %–50 % [3–6, 14]. In patients with congenital unilateral absence of the vas deferens (CUA VD), the incidence of ipsilateral CASV was 71 %–90 % [4, 6, 15] and contralateral CASV, 20 % [4]. In patients with ectopic vas deferens, agenesis of ipsilateral seminal vesicle would be inevitable [7, 9–13]. However, crossed ectopia of the vas may occur leaving the ipsilateral seminal vesicle at normal position [16]. The second patient in this report was found to have a crossed ectopic left vas emerging with a hypotrophic seminal vesicle, semen fructose test was mildly positive postoperatively, suggesting an under-functioning seminal vesicle. CASV may be associated with renal agenesis or other anomalies [1, 2], but the incidence was unclear. Although the patients may be in the condition of sterility, the testicular spermatogenesis is usually intact [5, 12], as in case 1.

Seminal vesicle secretion may promote sperm motility, increase stability of sperm chromatin and suppress the immune activity in the female reproductive tract to avoid rejection of spermatozoa and embryos that have antigens foreign to women [17]. No specific symptoms or signs are directly suggesting the diagnosis of seminal vesicle agenesis. CASV is frequently associated with CAVD or ectopia of the vas deferens, and patients with CASV may present with infertility, yet few cases can be found during routine ultrasonography or pelvic CT. Patients with ectopic vas into the Mullerian duct cyst can also manifest hematospermia, as the second case in our study. If the cyst is large enough, perineal and intrascrotal pain, dysuria, epididymitis, or urinary tract infection may reveal a cystic lesion in the prostate gland in patients with ectopic vas into the Mullerian duct cyst. In patients with CAVD, the scrotal vas is nonpalpable, and for most cases, the distal 2/3 segment of epididymis is absent leaving an enlarged and firm caput. There is still another kind of ectopia of the vas deferens where the vas terminated ectopically into the urinary tract, including bladder, posterior urethra, ureter or even kidney [7, 11–13]. Most cases of this condition were described in children and were frequently associated with congenital anorectal and other urogenital anomalies [7, 11, 13]. Our patients fell within a rare subgroup of people with ectopic vas–adults with infertility but without congenital renal, ureteral, or anorectal malformations.

CASV is not an independent condition. As stated before, it was always associated with CAVD or ipsilateral ectopia of the vas deferens. The clinical importance is usually underestimated and the diagnosis has been frequently neglected. Herein we recommended that in patients with infertility, vasal agenesis, low ejaculate volume, low semen pH and low semen fructose, it was mandatory to further inspect the appearance of the seminal vesicles. In patients with CAVD, vasography cannot be performed owing to ductal agenesis [14]. Transrectal ultrasonography (TRUS), CT and magnetic resonance imaging (MRI) were frequently used to detect seminal vesicle anomalies, but these imaging tools are incapable of diagnosing ectopia of the vas deferens. In patients with vasal ectopia, the invasive vasography is still necessary for confirmation. Abdominal ultrasonography, CT scans or excretory urography are helpful to determine the renal anomalies.

Congenital agenesis of the seminal vesicle is unreconstructable and requires no treatment in the subgroup of unilateral CASV with a potent contralateral ductal system. In patients with infertility, hematospermia, recurrent epididymitis, urogenital abnormalities, strategies concerning CAVD and ectopic vas deferens must be taken into consideration. Hall and Oates [15] have done well in treating the subgroup of male infertility patients who presented with CUA VD and contralateral ductal abnormalities. Corrective surgical options in this subset include vasoepididymostomy, transurethral resection of the ejaculatory duct and microscopic epididymal/testicular/vasal sperm aspiration [15]. The first case was within this subset, a transurethral unroofing of the Mullerian duct cyst was performed and semen analysis improved postoperatively.

The principles of treating ectopic vas deferens are to prevent epididymitis and urinary tract infection, to preserve fertility and to release the outlet obstruction [9]. Treatment of infertility due to vasal ectopia must be individualized [12]. Takahashi et al. [10] have reported a case with vasal ectopia into the Mullerian duct cyst. They
stated that open excision of the Mullerian duct cyst was preferred if the symptoms were severe or if other modes of therapy such as antibiotics had failed. The incisions include transabdominal, perineal and posterior approach [10]. We performed transurethral unroofing of the Mullerian duct cyst in two cases, because an extensive operation was not necessary. After operation, semen analysis improved in both cases and the symptoms of hematospermia in case 2 subsided.

Nowadays, assisted reproductive technology (ART) is available for patients with unreconstructable ductal obstruction. Microsurgical epididymal sperm aspiration or testicular sperm extraction combined with intracytoplasmic sperm injection (ICSI) have provided a viable treatment strategy for affected individuals [18–20]. Because testicular spermatogenesis is rarely impaired in patients with CASV, it is reasonable for them to undergo microsurgical epididymal sperm aspiration or testicular sperm extraction to obtain spermatozoa for ICSI.

Although ductal obstruction was eliminated and semen analysis was improved in our patients after operation, they remained infertile owing to the lack of functional seminal vesicles, sperm harvesting from epididymis or testis for ICSI is still necessary for them to start families. Both of the patients are recommended to our Hospital Center of Reproductive Medicine to receive ICSI.

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