Anomalies of the Wolffian Duct Derivatives Encountered at Radical Prostatectomy

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Abnormalities of wolffian duct derivatives are usually encountered in young subjects. We identified 4 instances of embryologic malformations of these structures in patients who underwent radical prostatectomy (RP). The first patient had unilateral renal agenesis and a seminal vesicle cyst identified preoperatively by computed tomography. The seminal vesicle cyst was removed during RP. The second patient had renal agenesis and an ectopic ureter entering the right seminal vesicle. These were treated with ureterectomy during RP. The third patient had unilateral duplication of the vas deferens of no clinical consequence. Finally, the fourth patient had left-sided absence of both the vas deferens and seminal vesicle. The anatomy of the lower pelvis is most accurately shown on magnetic resonance imaging. Lower urinary tract malformations are an uncommon occurrence in males. Surgeons who perform numerous RPs will, however, find additional urologic pathologies during RP that may require consideration and tailored management.
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Key words: Cyst • Prostate-specific antigen • Radical prostatectomy • Transrectal ultrasound • Wolffian duct derivatives

Patients evaluated and treated for clinically localized prostate cancer occasionally are found to have additional urologic pathologies. Transitional cell carcinoma of the bladder is the most common concomitant pathology in patients who undergo radical prostatectomy (RP), with reported incidences between 1.2% and 6.6%.[1] Although malformations of the genitourinary tract

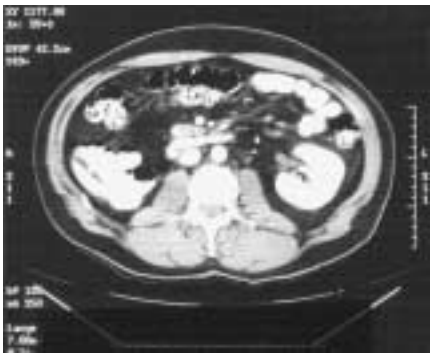

Figure 1. *Computed tomography scan from case 1 patient showing absence of the right kidney and collecting system with compensatory hypertrophy of the left kidney.*

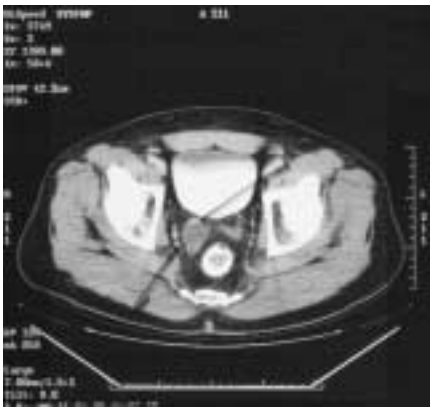

Figure 2. *Computed tomography scan from case 1 patient showing a large right seminal vesicle cyst.*

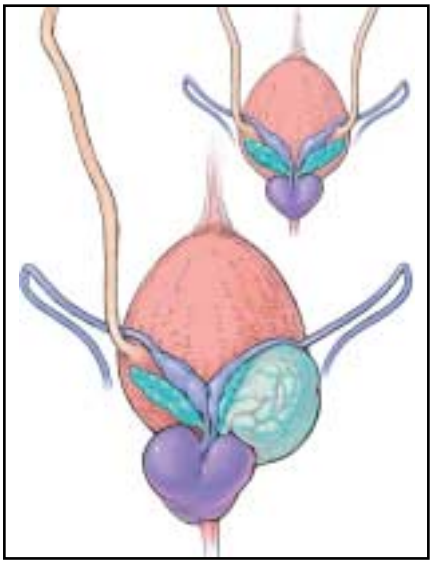

Figure 3. *Right-sided seminal vesicle cyst with congenital right renal and ureter agenesis.*

usually become apparent in childhood, they may remain silent until incidental detection during the evaluation and/or treatment of other pathologies detected in adulthood. We recently reported on the incidental detection of ureteroceles during the management of prostate cancer.[2] Herein, we report on the diagnosis, management, and embryologic origins of anomalies of wolffian duct derivatives such as renal agenesis, seminal vesicle cyst, ectopic ureter, and absence or duplication of vas deferens detected incidentally in patients who underwent RP for clinically localized prostate cancer.

Case Reports

Case 1

A 54-year-old white man was found to have a serum total prostate-specific antigen (PSA) level of 7.9 ng/mL during routine prostate cancer screening. He was referred to a community urologist, who noted a firm nodule in the left base during digital rectal examination (DRE). The patient was found to have Gleason score $4 + 4 = 8$ adenocarcinoma of the prostate in the left base, mid, and apical prostate on transrectal ultrasound (TRUS)–guided systematic sextant biopsies. Transrectal ultrasonography showed a hypoechoic mass with posterior acoustic enhancement arising in the right seminal vesicle measuring $2.0 \times 2.0 \times 1.9$ cm. Staging computed tomography (CT) and bone scans were both negative for metastatic disease but revealed absence of the right kidney and collecting system, compensatory hypertrophy of the left kidney (Figure 1), normal left collecting system, normal bladder, and a right seminal vesicle cyst (Figures 2 and 3). Magnetic resonance imaging (MRI) of the abdomen and pelvis showed right renal agenesis and a large mass, which measured 6 cm in greatest diameter, arising from the right seminal vesicle. The mass showed low T_1- and high T_2-weighted signal intensity compatible with a cyst, with a small central focus of high T_1- and T_2-weighted signal intensity, possibly representing hemorrhage and protein content.

The patient subsequently underwent RP and bilateral pelvic lymph node dissection (BPLND). Intraoperatively, a large seminal vesicle cyst was identified on the right side, and it was completely excised after careful identification and preservation of the neurovascular bundle. The final histopathologic report revealed pT3bN1M0, Gleason score $4 + 5 = 9$ adenocarcinoma, with established extracapsular extension along the left lateral lobe (Figure 4A), left-sided seminal vesicle involvement (Figure 4B), and negative surgical margins. The right seminal vesicle was cancer-free. It was cystic and elongated, with a tan-yellow, glistening lining. Unilateral sural nerve grafting was performed on the left side.[3] The postoperative recovery of the patient was uneventful, but he developed biochemical recurrence of his cancer within 18 months after surgery.

Case 2

A 62-year-old white man with lower urinary tract symptoms, including nocturia, who was otherwise healthy was found to have a serum total PSA level of 22.4 ng/mL during his routine yearly prostate cancer screening. His physical examination, including DRE, was normal. TRUS-guided sextant biopsy demonstrated a 32-cm^3 prostate with hypoechoic lesions in the left base and midprostate, which were found to involve a minute focus of Gleason score $3 + 3 = 6$ and a 2-mm focus of $3 + 2 = 5$ prostate adenocarcinoma, respectively. On preoperative staging CT, the patient was

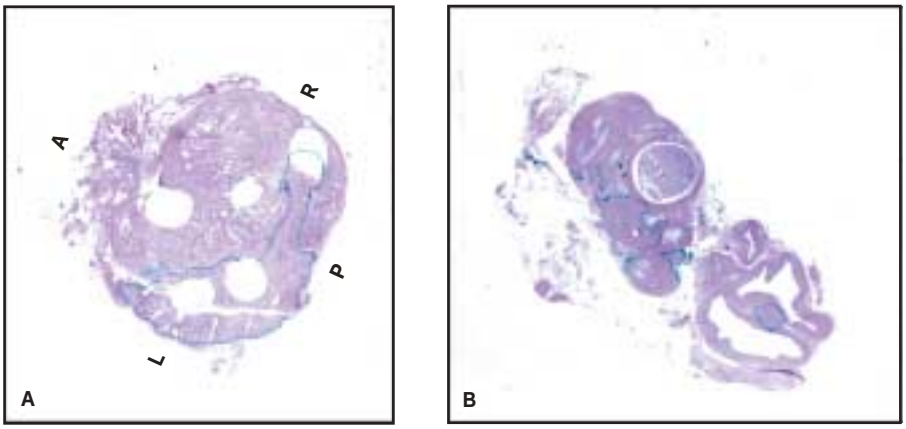

Figure 4. *Whole-mount 5-mm step-sectioned histologic slides from the prostate of case 1 patient showing* (A) *established extracapsular extension along the left lateral lobe and* (B) *left-sided seminal vesicle involvement.*

found to have right renal agenesis with a dilated ipsilateral ureter ending in the seminal vesicle. The contralateral kidney was hypertrophied but had a normal collecting system.

Intraoperatively, the patient was found to have a hydronephrotic ectopic ureter approximately 1 cm in diameter that ended blindly in the right seminal vesicle. Opening of the bladder revealed that the right hemitrigone and ureteral orifice were absent. The right ureter was transected at the seminal vesicle. Attempts to pass a catheter into the right ureter pointing to the seminal vesicle were unsuccessful. A right retrograde pyelogram revealed that the right ureter ended blindly in the retroperitoneum. A complete ureterectomy was performed in conjunction with the RP. The bladder neck was reconstructed in standard fashion. The patient tolerated the procedure well and experienced mild postoperative numbness in the groin, which resolved within several days. The remainder of the patient's recovery was uneventful. The patient was cancer-free at last follow-up.

Case 3

A 58-year-old previously healthy white man was referred to us because of a serum total PSA of 8.8 ng/mL and a percent free PSA ratio of 8%. His medical history and physical examination, including DRE, were unremarkable. TRUS revealed a 20-cm^3 prostate with hypoechoic lesions in bilateral apical peripheral zones. TRUS-guided systematic 12-core biopsy revealed small foci of Gleason score 4 + 4 = 8 adenocarcinoma in bilateral apexes.

The patient underwent RP and BPLND. Intraoperatively, a duplicated vas deferens was incidentally identified on the right side (Figure 5). After the bladder neck was opened, placement of a urethral stent in the right urethral orifice confirmed that the additional tubular structure was a duplicated vas deferens and not an ectopic ureter; therefore, it was left in situ. The contralateral anatomy was normal. The remainder of the procedure was performed without complications. The patient had an uneventful postoperative convalescence.

Case 4

A 53-year-old white man underwent a TRUS-guided systematic sextant biopsy at another institution because of a suspicious DRE and a preoperative serum total PSA level of 3.8 ng/mL. He was found to have Gleason score 3 + 3 = 6 prostate adenocarcinoma. Physical examination and metastatic work-up were negative. During TRUS-guided staging biopsy at our institution, he was found to be missing the left seminal vesicle.

The patient elected to undergo RP and BPLND. Intraoperatively, absence of the left seminal vesicle was confirmed along with absence of the left vas deferens. The right vas deferens and seminal vesicle were intact. There was no left ureteral orifice. The bladder neck was reconstructed in standard fashion. There was no complication, and the patient's recovery was uneventful. Final histopathology revealed Gleason score 4 + 3 = 7 pT3aN0Mx adenocarcinoma with focal extraprostatic extension but without surgical margin or right seminal vesicle involvement.

Discussion

One third of congenital anomalies are located within the kidneys or the urinary tract. These anomalies are uncommon, however, with a reported prevalence of 0.1%-1% in newborns.[3-5] The majority of these anomalies do not cause clinical problems and thus remain undetected. Unilateral renal agenesis is found in 1 of 1100 autopsies,[6] whereas clinically significant presentation may occur in only 1 in 1500.[7] Unilateral renal agenesis is often associated with other anomalies. Approximately 30%-40% of men with unilateral renal agenesis have been reported to have anomalies of the urogenital tract or other systems.

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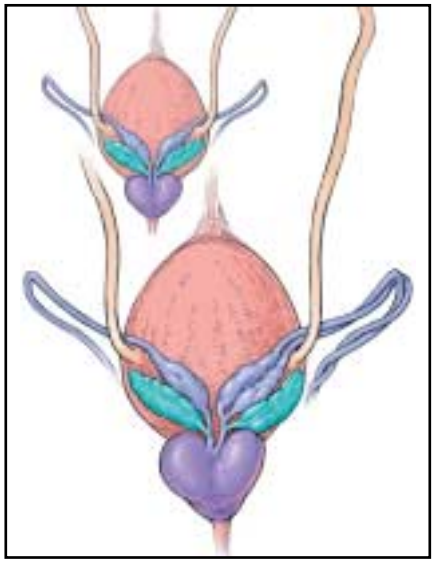


Figure 5. *Right-sided true duplication of the vas deferens.*

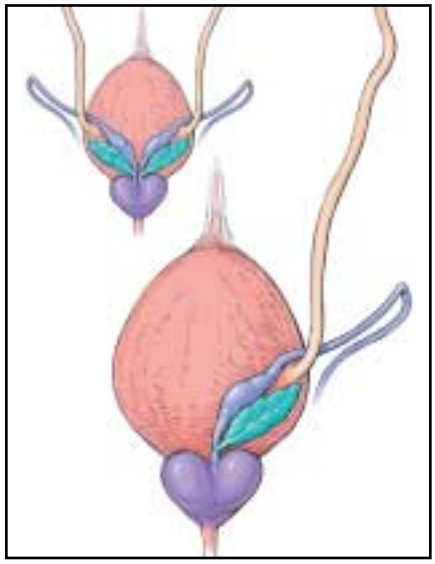
Unilateral renal agenesis may be associated with other congenital anomalies, such as cardiovascular (30%), gastrointestinal (25%), and/or musculoskeletal (14%) anomalies.[8] Genital abnormalities and cystic masses within the pelvis also may occur. During a renal sonographic mass screening of 280,000 children in Taipei, Taiwan, Sheih and colleagues[9] found congenital renal agenesis in 236 children (119 boys and 117 girls). Six of the 119 boys had a concomitant ipsilateral seminal vesicle cyst, and 7 of the 117 girls had a concomitant ipsilateral Gartner cyst, the embryologic equivalent in the female.

In 1914, Zinner[10] described the first case of a seminal vesicle cyst (Figure 3) combined with ipsilateral renal agenesis. Since then, approximately 135 cases of seminal vesicle cysts have been reported; approximately 52 were associated with renal agenesis.[11] Seminal vesicle cysts may be congenital or acquired. Present since birth, congenital cysts develop and become symptomatic in the second to third decade, during the period of greatest sexual or reproductive activity.[12] Accumulation of secretions in the gland resulting from insufficient drainage, which is associated with atresia of the ejaculatory ducts, causes distention, leading to formation of a cyst. The cysts usually are unilateral, with no predilection for either side. Acquired cysts are often bilateral and are seen in an older group with a history of chronic prostatitis or prostate surgery. Seminal vesicle cysts smaller than 5 cm may remain asymptomatic and are usually discovered incidentally as a palpable abdominal mass or on DRE as a palpable fluctuant mass arising from the superior aspect of the prostate gland. In one study, 79% of seminal vesicle cysts were palpable on DRE.[11] These cysts may also be associated with symptoms related to bladder irritation and obstruction, such as dysuria, frequency, hematuria, urinary tract infections, perineal pain, pain following ejaculation, scrotal pain and/or symptoms of epididymitis and prostatitis.[11,13,14] Cysts of the seminal vesicle larger than 12 cm have been termed "giant" cysts and often are present with symptoms of bladder and/or colonic obstruction because of mass effect.[13]

Seminal vesicle cysts are commonly associated with ipsilateral renal agenesis because of their common embryologic origin. With normal development, in the fourth embryonal week, the ureteric bud sprouts from the dorsal aspect of the distal mesonephric (wolffian) duct and extends dorsocranially into the central part of the metanephric blastema, which induces the differentiation of the metanephric blastema into the nephron. The ureteric bud differentiates into the ureter and the collecting duct system of the kidney, whereas the mesonephric duct differentiates into hemitrigone, bladder neck, urethra (proximal to the external sphincter), seminal vesicle, vas deferens, epididymis, paradidymis, and appendix epididymis. Complete failure of the mesonephric duct results in absence of the ipsilateral kidney, ureter, hemitrigone, vas deferens, and seminal vesicle (Figure 6). Failure of the ureteral bud to develop and meet the metanephric blastema will lead to ipsilateral renal agenesis or dysplasia with atresia of the ejaculatory duct, resulting in obstruction of the seminal vesicle and formation of cysts.[15] If the ureteral bud arises in a more cephalic position, delayed absorption of the caudal mesonephric duct will result in the distal ureteral bud emptying into mesonephric derivatives. Tumors of the seminal vesicle appear to be extremely rare, with only 2 reported cases of papillary carcinoma in a seminal vesicle cyst.[16]

Ectopic ureters entering seminal vesicle cysts (Figure 7) associated with ipsilateral renal agenesis are uncommon.[17] Ectopic ureters are more commonly detected in females than in males, with an estimated 5:1 ratio. Although ectopic ureters are usually associated with duplication of the drainage system of the involved kidney in females, only one fourth of

Figure 6. *Left-sided congenital absence of the vas deferens, seminal vesicle, ureter, and kidney.*



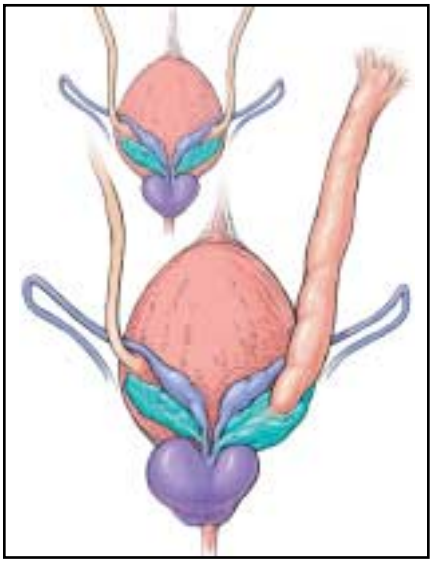


Figure 7. *Congenital right renal agenesis and large right-sided ectopic ureter with proximal hydronephrosis and distal blind-ended entry into the right seminal vesicle.*

males present with a duplicated system. If the ureteral bud develops more cephalad on the mesonephric duct, it will not achieve an independent opening into the urinary triad. The ureter remains attached to the distal part of the mesonephric duct that differentiates into seminal vesicle and ejaculator duct.[18] Often, a rudimentary renal unit is detected on the involved side and its ureter ends in the ipsilateral vas deferens or seminal vesicle.[19] Enlarged ectopic ureters are normally diagnosed by ultrasound for work-up of pyuria and/or acute pyelonephritis in neonates, and epididymitis, flank pain, and/or palpable seminal vesicle in prepubertal adolescent boys.[20] Postpubertal males may experience prostatitis, pain on ejaculation, and possibly infertility.

Duplication of the vas (ductus) deferens (Figure 5) is a rare condition, with only a few cases reported in the literature. The embryologic etiology of this maldevelopment is incompletely understood. It has been suggested that the duplication of the vas deferens may be caused by duplication of the fetal mesonephric system. According to Vohra and Morgentaler,[21] the term *true duplication* of the vas deferens should be restricted to cases in which a duplicate vas is identified within the spermatic cord and should be distinguished from the term *double vas deferens*, which is used in several case reports in the literature to describe an ectopic ureter draining into the seminal vesicle.[18,21] Gibbons and colleagues[22] suggested that abnormalities of the vas deferens and ureter are the result of maldevelopment of the proximal vas precursor, a segment next to the mesonephric duct that differentiates into the vas deferens and seminal vesicles.[22]

Several imaging techniques have been used in the evaluation and differentiation of pelvic anomalies. Sonography can detect and confirm the cystic nature of pelvic masses but is a poor choice for imaging of the upper tract. We identified the seminal vesicle cysts in the first and fourth case using TRUS of the prostate. Furthermore, as we reported previously, ureteroceles may also be identified incidentally during TRUS.[2] Intravenous urography can show ipsilateral renal dysgenesis and an abnormal appearance of the excretory system. However, seminal vesicle cysts cannot be visualized. CT scans can accurately show renal anomalies and define pelvic anatomy. The ideal imaging study is MRI, however, because it defines abdominal and pelvic anatomy, demonstrating the relationships among different pelvic structures, and differentiates malformations of the pelvis with high accuracy. Currently, though, only patients with high-grade prostate cancer or those with markedly high PSA levels undergo additional imaging, such as CT or

It has been suggested that the duplication of the vas deferens may be caused by duplication of the fetal mesonephric system.

Main Points

- Although malformations of the genitourinary tract usually become apparent in childhood, they may remain silent until incidental detection during the evaluation and/or treatment of other pathologies detected in adulthood.
- One third of congenital anomalies are located within the kidneys or the urinary tract. The majority of these anomalies do not cause clinical problems and thus remain undetected.
- Although rare, urologic abnormalities are a possibility to be considered in planning treatment for men with prostate cancer.
- Computed tomography scans can accurately show renal anomalies and define pelvic anatomy. The ideal study is magnetic resonance imaging, however, because it defines abdominal and pelvic anatomy, demonstrating the relationships among different pelvic structures, and differentiates malformations of the pelvis with high accuracy.

MRI. Vesiculodeferentography and cystoscopy are invasive procedures and usually are not performed.

Conclusion

Although rare, urologic abnormalities are a possibility to be considered in planning treatment for men with prostate cancer. We report on 4 asymptomatic men who presented for work-up and treatment of their prostate cancer and were found to have incidental urologic abnormalities secondary to embryologic malformation. The variety of asymptomatic malformations that may be found preoperatively by radiographic imaging either in the work-up for initial diagnosis or at the time of staging of prostate cancer prompt tailored management strategies for these patients. For example, preoperative identification of the seminal vesicle cyst in case 1 allowed for the surgeon to plan ipsilateral nerve-sparing RP. In case 3, on the other hand, the duplicated vas deferens was not diagnosed preoperatively because of the lack of an upper tract. Preoperative identification of the abnormality would have prompted a functional study of the upper tract to rule out a source of infection or obstruction. ■

Disclosure

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