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A case of congenital unilateral absence of the vas deferens

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Background: Congenital unilateral absence of the vas deferens occurs in 0.5%–1.0% of males. It has been associated with various genitourinary abnormalities, including renal agenesis. We report a case of congenital unilateral absence of the vas deferens found incidentally during vasectomy in a patient with known unilateral renal agenesis.

Case presentation: A 24-year-old male presented to our urology clinic requesting vasectomy. His past history was significant for left renal agenesis. Following successful right vasectomy, several attempts to locate the left vas deferens were unsuccessful. We diagnosed congenital unilateral absence of the vas deferens. Follow-up semen analysis showed azoospermia.

Conclusion: As vasectomies are increasingly performed in outpatient settings, it is imperative that physicians be aware of this condition, which can be recognized by a simple physical exam. Recognition could prevent unnecessary surgery and prompt providers to investigate for associated abnormalities.

Keywords: vas deferens, embryology, abnormalities, surgery

Background

Congenital unilateral absence of the vas deferens (CUAVD) was first described by John Hunter in 1737.¹ Reverdin in 1870 described the association between vas deferens agenesis and ipsilateral renal agenesis.² CUAVD is an uncommon entity with a reported prevalence range of 0.5%–1% in the male population. It has been associated with renal agenesis and a variety of other anomalies. As vasectomies are being increasingly performed by family medicine physicians they may encounter this condition more frequently. Awareness of this condition would help uncover coexisting anomalies.³

We present a case of CUAVD which was found incidentally during a vasectomy in a patient with ipsilateral renal agenesis.

Case presentation

A 24-year-old male presented to our urology clinic requesting vasectomy. The patient was married and had fathered one child with his wife. A detailed pre-operative history showed that the patient had left renal agenesis. There was no documented evaluation for any other concomitant anomalies of the genitourinary system. Written consent was obtained. During the procedure the patient was locally anesthetized with 1% xylocaine. His right vas deferens was palpated; a small transverse incision was made in the upper midline region of the scrotum. The right vas deferens was isolated, doubly clamped and the intervening segment excised and sent for pathology, which subsequently confirmed

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it as the vas deferens. After hemostasis was achieved the left hemiscrotum was palpated, however the vas deferens was not identified. A structure thought to be the atrophic vas deferens was isolated, doubly clamped and the intervening segment sent for pathology; however, frozen section revealed it to be a blood vessel. After suturing the blood vessel and closing the incision, an incision was made 2 cm away where the left vas deferens was thought to be palpated; however, it turned out to be the stitches placed on the previously mentioned blood vessel. The incisions were closed and bandages were applied. The patient was diagnosed with unilateral absence of the vas deferens. He was discharged in stable condition. He returned for follow-up two months later, at which time his incisions had healed and he was doing well. The patient's post-operative semen analysis showed azoospermia. The patient was not tested for cystic fibrosis trans membrane conductance regulator (CFTR) gene mutation as he had completed his family and there was no risk of further genetic transmission.

Discussion

CUAVD is a rare entity and has an incidence of 0.5%–1% in the male population as described above.³ It is commonly discovered either during an evaluation for infertility or during a vasectomy.⁴ About 0.06%–0.8% of the patients undergoing vasectomy have CUAVD.³ CUAVD occurs twice as commonly on the left as compared to the right side. It is divided into two subgroups based on the fertility status. Patients who were fertile had a high incidence of ipsilateral renal agenesis. Patients who were not fertile had a partially obstructed contralateral vas deferens but normal upper urinary tracts.³

Renal agenesis is more commonly associated with unilateral vasal agenesis (73.7%) compared to the bilateral form (11.8%).³ CUAVD occurring with renal agenesis is due to an intrinsic Wolffian duct defect. In the embryo, the vas deferens arises from the Wolffian duct, which connects the mesonephros and the urogenital sinus. The ureteral bud arises from the Wolffian duct as it curves to enter the cloaca. The penetration of the metanephrogenic blastema by the ureteric bud induces the development of the kidney. Any interruption of this process before the complete separation of the Wolffian duct and ureteric bud can result in renal agenesis and CUAVD whereas interruption in the development of the Wolffian duct after the separation may lead to an isolated unilateral absence of the vas deferens.²

Other renal anomalies associated with CUAVD are malrotation of the solitary kidney, multicystic kidney, ectopic

kidney, and horseshoe kidney.⁵ Anomalies of the seminal vesicles, ejaculatory ducts, cryptorchidism, and inguinal hernia have also been reported in association with CUAVD.⁴ Rare cases of vesicoureteric reflux and ureterovesical obstruction in the contralateral kidney have also been described.³ Ipsilateral agenesis of the adrenal glands has also been reported occurring with renal agenesis and absence of the vas deferens.⁶

About 1%–2% of males investigated for infertility have congenital vasal agenesis.³ Most of these cases are due to bilateral vas agenesis (1%–6%). Only 0.4% of male infertility cases have been attributed to CUAVD. The infertility in CUAVD patients is often due to obstruction of the contralateral vas deferens.³ Casals et al showed that 38% of congenital unilateral absence of the vas deferens cases are associated with mutations in the CFTR gene.⁷ About 45% of these mutations were specific to congenital absence of the vas deferens (CAVD) and were not found in cystic fibrosis patients. The OLA/PCR mutation detection system which detects 31 different cystic fibrosis mutations identified only 26% of the CFTR mutations in CAVD patients. This underlines the importance of an extensive CFTR gene analysis in these patients. Previous studies had found low association between renal agenesis and CFTR mutations. According to the authors, the extensive CFTR gene analysis done in place of the routine screening might have been responsible for the increased detection of CFTR mutations in CUAVD patients.⁷ CFTR gene analysis should be offered to CAVD patients with infertility planning to have children using assisted reproductive techniques as there is a risk in the offspring to have cystic fibrosis or related diseases.⁸

CAVD may be tentatively diagnosed in the absence of the firm, round, sliding linguini-like vas deferens that sits in continuation with the epididymis and can be followed to the external inguinal ring on palpation.³ Vasography or transrectal ultrasonography can also be utilized to visualize a missing segment of this structure. However, before diagnosing CAVD, a scrotal exploration under general anesthesia is recommended.⁴ As shown by Weiss, considerable delay (4.3 years on average) can occur before CAVD is diagnosed.⁵ This can potentially lead to increased mortality and morbidity associated with renal anomalies. Therefore, palpating the vas deferens should be a part of the routine physical exam in males. Patients diagnosed with CAVD should be encouraged to undergo an abdominal ultrasound study to detect any other coexisting abnormalities.^{3,4} Conversely, patients with known renal agenesis should also be evaluated for anomalies of the vas deferens.

Conclusion

As vasectomy is the most common method of sterilization, it is essential that providers be aware of this condition and its associated abnormalities. Palpation of the vas deferens should be included as a part of the routine physical exam in males. Patients diagnosed with CUAVD should undergo renal imaging for detection of any other anomalies.

Disclosure

The authors report no conflicts of interest in this work.

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