# CONGENITAL ABSENCE OF BILATERAL SEMINAL VESICLES AND VAS DEFERENS WITH RIGHT RENAL AGENESIS AND LEFT ECTOPIC KIDNEY

Sadia Babar, Umar Amin, Rashid Nazir, Imaad-ur-rehman, Atif Rana

Department of Radiology and Imaging, Shifa International Hospital, Islamabad, Pakistan

PJR January - March 2011; 21(1): 31-33

## Introduction

Congenital absence of seminal vesicles and vas deferens is an uncommon anomaly that contributes to male sterility. Bilateral SV agenesis is associated with mutations in the cystic fibrosis transmembrane conductance regulator gene in 64%–73% of cases and may be related to luminal obstruction by thick secretions. It is also frequently associated with bilateral agenesis of the VD. Patients usually have normal kidneys. We present a case of bilateral agenesis of seminal vesicles and vas deferens with absent right kidney and pelvic left kidney.

# Case Report \_\_

A 31-year-old male patient married for 10 years presented with complaint of infertility. His wife had undergone relevant workup and her reports were normal. Semen analysis of the patient yielded azoospermia with low volume of 1ml and low pH of 6.5. Semen fructose was negative and erythtocytes were absent. Hormonal analysis showed normal leutinizing hormone (LH), follicle stimulating hormone (FSH) and testosterone levels. His scrotal ultrasound was normal and FNAC of aspirate from both testes showed normal spermatogenesis suggestive of obstructive azoospermia. Transrectal ultrasound was performed in our department, which showed absence of bilateral seminal vesicles and vas deferens. Prostate gland was normal (Fig.1). He also had complaints of burning and difficulty in micturition. Transabdominal

Correspondence: Dr. Sadia Babar Department of Radiology and Imaging, Shifa International Hospital, Islamabad, Pakistan Email: sadia.babar@ymail.com



Figure 1: Transrectal ultrasound scan showing absent bilateral seminal vesicles and vas deferens. S= Expected location of seminal vesicles, White arrow= Normal prostate

ultrasound performed for evaluation of genito- urinary tract, showed right sided absent kidney with ectopic left kidney in left iliac fossa adjacent to the urinary bladder (Fig.2).

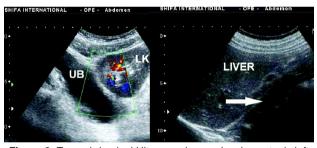


Figure 2: Transabdominal Ultrasound scan showing ectopic left kidney placed adjacent to the urinary bladder. White arrow shows absent right kidney. UB= Urinary Bladder, LK= Left kidney

### **Discussion**

Anomalies of the seminal vesicles can occur as abnormalities of number (agenesis, fusion, duplication), maturation (hypoplastic), position (ectopia) and structure (diverticulum, cyst, communication with the ureter).<sup>2</sup> Among all seminal vesicle cysts are commonest. Their significance lies in late diagnosis and association with other mesonephric duct anomalies.

Mesonephric duct differentiates into the appendix of the epididymis, paradidymis, vas deferens, ejaculatory duct, seminal vesicle, and hemitrigone of bladder. The ureteral bud arises off the dorsal aspect of the distal mesonephric duct and extends in a dorsocranial fashion to meet and induce differentiation of the metanephric blastema, which will form the adult definitive kidney.<sup>3</sup> Complete failure of the mesonephric duct to develop will result in failure of development of its derivatives. If insult occurs prior to seven weeks of gestation in utero i.e before the development and induction by the ureteral bud, will lead to renal agenesis.<sup>4</sup> During fetal development, ureteral buds first appear inside the pelvis, near the bladder and then ascent to their normal position. Failure of metanephros to ascent leads to ectopic kidney.

Congenital absence of bilateral seminal vesicles is frequently associated with congenital bilateral absence or ectopia of vas deferens<sup>5</sup> with 64 -73% of cases having mutations in the Cystic fibrosis transmembrane receptor CFTR gene. A study also states 80% of CFTR mutations in congenital bilateral absence of vas deferens CBAVD patients and this may contribute to the close association between these two entities.<sup>6</sup> The same study shows renal agenesis in 5.4 percent of patients with CBAVD. The maldevelopment of kidneys though is not explainable in these cases. It is postulated that renal anomalies are present in patients with CBAVD and CASV who do not have mutations in CFTR gene and may represent a different genetic basis.<sup>7,8</sup> Thus, renal agenesis with opposite ectopic kidney is a rare finding in patients with CABVD and CASV.

Seminal vesicles and prostate contribute 90 percent of fluid in ejaculate and seminal vesicles also contribute in making the ejaculate alkaline. These patients usually present late with complaints of infertility. Approach to diagnosis is usually made by semen analysis according to protocol set by WHO criteria. Testicular biopsy and aspirate analysis is considered mandatory in patients with normal hormonal levels, however in some cases vasography may also be considered. Transrectal ultrasound provides good visualization of prostate and seminal vesicles. CT and MRI are the newer investigations being carried out for diagnosis.9 Unilateral renal agenesis also does not present with symptoms until and unless there is some pathology in the contralateral kidney which in our case was ectopic causing dysuria. Transabdominal ultrasound, IVU and CT pyelogram can be done for evaluation of urinary tract in these patients.

Congenital agenesis of seminal vesicle is not surgically correctable. Treatment options include ICSI i.e. sperm retrieval by TESE (Testicular sperm extraction) and implantation by IVF.<sup>10</sup>

# **Abbreviations**

SV = Seminal vesicles
VD = Vas Deferens

FNAC = Fine needle aspiration cytology

CBAVD = Congenital bilateral absence of vas deferens
CASV = Congenital absence of Seminal vesicles

CT = Computed TomographyMRI = Magnetic resonance Imaging

# References

- Bohyun Kim, Akira Kawashima, Jeong-Ah Ryu, Naoki Takahashi, Robert P. Hartman, and Bernard F. King, Jr. Imaging of the Seminal Vesicle and Vas Deferens. Radiographics July-August 2009; 29: 1105-21.
- 2. Wu HF, Qiao D, Qian LX, et al. Congenital agenesis of seminal vesicle. Asian J Androl 2005;7: 449-52.
- 3. Livingston L, Larsen CR. Seminal vesicle cyst with ipsilateral renal agenesis. AJR 2000;**175**: 177-80.
- S. S. Arora, R. S. Breiman, E. M. Webb, A. C. Westphalen, B. M. Yeh, and F. V. Coakley. CT and MRI of Congenital Anomalies of the Seminal Vesicles.Am. J. Roentgenol., July 1, 2007; 189(1): 130-5.
- R. S. Narlawar, V. Hanchate, A. Raut, P. Hira, A. Nagar, and N. G. Chaubal. Renal Agenesis and Seminal Vesicle Cyst. JUM February 2003;22: 225-8.
- 6. Teresa Casals, Lluís Bassas, Susanna Egozcue, Maria D. Ramos, Javier Giménez, Ana Segura, Ferran Garcia, Marta Carrera, Sara Larriba, Joaquim Sarquella, and Xavier Estivill. Heterogeneity for mutations in the CFTR gene and clinical correlations

- in patients with congenital absence of the vas deferens. Hum. Reprod. (2000); **15(7)**: 1476-83.
- T.J. McCallum, J.M. Milunsky, R. Munarriz, R. Carson, H. Sadeghi-Nejad, and R.D. Oates. Unilateral renal agenesis associated with congenital bilateral absence of the vas deferens: phenotypic findings and genetic considerations. Hum. Reprod. (2001);16(2): 282-8.
- 8. Ramin Radpour, Hamid Gourabi, Mohammad Ali Sadighi Gilani, Ahmad Vosough Dizaj. Correlation Between CFTR Gene Mutations in Iranian Men With Congenital Absence of the Vas Deferens and Anatomical Genital Phenotype. Journal of Andrology, January/February 2008; 29(1).
- Nitin P Ghonge, Bharat Aggarwal, and Amit Kumar Sahu. Zinner syndrome: A unique triad of mesonephric duct abnormalities as an unusual cause of urinary symptoms in late adolescence. Indian J Urol. 2010 Jul-Sep; 26(3): 444.
- ID Sharlip, PP Urology, J Jarow, AM Belker et al. Report on management of Azoospermia. An AUA Best Practice Policy and ASRM Practice Committee Report