CONGENITAL BILATERAL APLASIA OF VAS DEFERENS (CBAVD): A REMINDER TO CLINICIANS

Koh PS, Shanggar K, Razack AH, Lee G.

Department of Surgery, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur.

ABSTRACT

Male factor infertility which accounts for 30-50% of infertility is a major problem faced by married couples. Congenital absence of the vas deferens, though uncommon, remains the most common abnormality seen in extratesticular ductal and ejaculatory system, accounting for 1-2% of male infertility. It may be unilateral or bilateral. Association with renal abnormality has also been reported with congenital absence of vas deferens (1). The patients are asymptomatic and the congenital abnormality is usually detected when investigation for infertility is carried out. We present a case of an unusual presentation of congenital bilateral absence of vas deferens (CBAVD). *(JUMMEC 2008; 11(2):89-90)*

KEYWORDS: absent vas deferens, renal agenesis, infertility

Introduction

Male factor infertility which accounts for 30-50% of infertility, is a major problem faced by any couple. Congenital absence of the vas deferens though uncommon, remains the most common abnormality seen in extratesticular ductal and ejaculatory system abnormalities accounting for 1-2% of male infertility. It may be unilateral or bilateral. It was first described by John Hunter in 1737 in a cadaver. We report a case of a young man who is not considering conception with congenital bilateral aplasia of vas deferens (CBAVD).

Case Report

A 20-year-old single man with no previous illnesses was admitted with 2 years' history of reduction in ejaculatory volume and reduction in consistency of the ejaculatory content. Bilateral testes and epididymis examinations were normal, but bilateral vas deferens was not palpable. The semen analysis revealed 1.7 ml of ejaculate, which was acidic in nature (pH 6.8) and demonstrated azoospermia. His hormonal profile, including FSH, LH, prolactin and teststosterone were normal. An ultrasound of the scrotum confirmed the absence of bilateral vas deferens with normal testes and epididymis. Intravenous urogram (IVU) also showed an absent left kidney (Figure 1). The patient was reassured and advised to proceed with further investigation and potential in-vitro fertilization (IVF) when considering conception.

Discussion

Congenital bilateral absence of vas deferens is an uncommon anomaly contributing to 1-2% of male infertility (1, 2). It was first described by John Hunter in 1737 in a cadaver. Absence of vas may be unilateral or bilateral (3, 4). Absence of vas deferens can be associated with various congenital anomalies such as renal anomalies (i.e. agenesis, malrotation, ectopia, fusion, or polycystic disease), inguinal hernias, varicocoeles or cryptorchidism. Renal agenesis in association with absence of vas deferens has been reported to be around 10-30% (unilateral and bilateral absence of vas deferens), though one report claimed up to 80-90% of unilateral absence of vas deferens is associated with ipsilateral renal agenesis (2, 3). Congenital absence of vas deferens has also been found to be associated with cystic fibrosis, an autosomal recessive disorder. Genetic screening for cystic fibrosis transmembrane conductance regulator (CFTR) is advised in those with congenital absence of vas deferens. The role of CFTR screening in patients with renal agenesis is debatable, as various reports by Schelegel et al, Augarten et al and

Correspondence: Shanggar Kuppusamy Department of Surgery Faculty of Medicine University of Malaya, 50603 Kuala Lumpur Email: drshanggar@um.edu.my



Figure 1: Intravenous Urogram showing absence of the left kidney

Dumur *et al* observed that there was no detectable CFTR mutations in patients with renal agenesis associated with congenital absence of vas deferens. Thus, it was suggested that those with congenital absence of vas deferens with renal agenesis do not usually have a genital form of cystic fibrosis (2, 5, 6). The currently recommended management of couples with infertility owing to CBAVD is sperm retrieval combined with IVF using intracytoplasmic sperm injection (ICSI) after appropriate genetic testing and counseling of the couple regarding the risk of cystic fibrosis. A majority of the patients with CBAVD are asymptomatic and the conhenital abnormality is usually detected present during the investigation for infertility. We present a case of an unusual presentation of a single man with symptoms of reduction in semen volume and consistency, as the first presentation of CBAVD.

References

- 1. Maria Rosa Maduro and Dolores J Lamb. Under standing the new genetics of male infertility. *J Urol,* Nov 2002; Vol 168: 2197-2205.
- Schlegel, Peter N; Shin, David; Goldstein, Marc. Urogenital anomalies in men with congenital absence of the vas deferens. *J Urol* May 1996; Vol 155: 1644-1648.
- 3. ZAJ Khan, JR Novell. A missing vas: A case report. J R Soc Med 2001; 94: 582-583.
- Hunter, J. On the glands called vesiculae seminales.
 In: Palmer JF, *Complete works*. London: Longman, Reese, Orm, Brown, Green & Longman, 1737; 4: 23.
- 5. Augarten, A; Yahav, Y; *et al.* Congenital bilateral absence of vas deferens in the absence of cystic fibrosis. *Lancet* 1994; 344: 1473.
- 6. Dumur, V; Gervais, R; Rigot, JM; *et al.* Congenital bilateral absence of vas deferens in absence of cystic fibrosis. *Lancet* 1995; 345: 200.