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Recommended Citation

Moazzam, M., Biyabani, S. R., Jadoon, C. K. (2003). Isolated congenital bilateral absence of vas deferens (CBAVD): A rare anomaly and diagnostic dilemma. *Journal of the Pakistan Medical Association*, 53(8), 372-373.

Available at: https://ecommons.aku.edu/pakistan_fhs_mc_surg_urol/118

Isolated Congenital Bilateral absence of Vas deferens (CBAVD): a rare anomaly and diagnostic dilemma

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Introduction

Congenital bilateral absence of Vas deferens (CBAVD) is an uncommon anomaly that contributes to male factor infertility. Various congenital abnormalities can occur in the course of mesonephric duct development which poses difficulties for diagnosis and management of such patients.

Case Report

Case 1

A 32-year old male shopkeeper presented with a 8-year history of primary infertility. History and clinical examination were unremarkable. Two urologists examined the patient separately and both had reported clinically palpable vas deferens. Seminal examination on two occasions, showed low volume (less than 1.5ml) fructose-positive azoospermia. Hormonal profile including serum testosterone, prolactin, LH and FSH were normal. Scrotal exploration + vaso-epididymal anastomosis was planned. At exploration, vas deferens was found to be bilaterally absent, epididymal head and body were dilated along with blindly ending tail. Epididymal aspirate showed numerous normally looking sperms. A transrectal ultrasound scan (TRUS) (Figure) showed bilateral presence of seminal vesicle. Subsequently the patient was offered epididymal aspiration and intracytoplasmic sperm insemination.

Case 2

A 25-years old male presented with primary infertility, for 4 years. History and clinical examination were unremarkable. He was examined by two urologists separately and both had documented bilateral clinically palpable vas deferens. Three serial seminal examinations showed low volume ejaculate and fructose- positive azoospermia. Hormonal profile was normal. A TRUS showed bilateral presence of seminal vesicles. Scrotal exploration + vaso-epididymal anastomosis was planned. On exploration, he was found to have bilaterally absent vas deferens. Epididymis was found to be dilated along with blindly ending tail. Epididymal aspirate showed numerous normal looking sperms.

Discussion

The seminal tract develops from the mesonephric duct which is a paired structure draining the mesonephric tubules in to the cloaca. Ureteric bud arises from the mesonephric duct at 4th week of gestation just proximal to the cloaca and subsequently it separates from the mesonephric duct at 7th week of gestation. After separation of ureteric bud, mesonephric duct gives origin to ejaculatory duct, seminal vesicles and ampulla of vas deferens from caudal part, vas deferens from its middle part and distal 2/3rd of

epididymis from its cranial part.¹ Common associated congenital abnormalities found in CBAVD include cryptorchidism and patent processus vaginalis.²

Previous investigators have suggested that with congenital absence of vas deferens, seminal vesicles and ejaculatory ducts are also absent. However in this sub-set of patients the entire mesonephric duct development was not abnormal as shown by presence of different parts of seminal tract.

About 10% of infertile males are diagnosed to have obstruction of the seminal tract which may be congenital or secondary to inflammatory events or surgery. Most frequent obstructive malformation of the seminal tract is bilateral congenital absence of vas deferens.² Almost all of these patients have bilaterally absent seminal vesicles and if present they are usually hypoplastic and non functional along with absence of ejaculatory ducts. These patients have low volume ejaculate (less than 1.5ml), absence of fructose in the semen and azoospermia as shown by the some of the studies in these patients.

Pre-operative clinical examinations are misleading in cases of vas aplasia, so surgical exploration is necessary in these patients.³ In our patients as well, clinical examination was not helpful. Ewa Kuligowka et al had studied the incidence of different concomitant congenital abnormalities of seminal tract along with CBAVD using transrectal ultrasound in patients with low volume ejaculate and azoospermia. Ninety six (34%), out of 276 patients had CBAVD. All of these patients were found to have bilaterally absent ejaculatory ducts and only one patient was found to have bilaterally present seminal vesicles.⁴ Both of our patients had TRUS but in both patients radiologist was unable to localize the vas deferens but reported the presence of seminal vesicles bilaterally.

Mac Goldstein et al studied the presence of seminal vesicles in patients with CBAVD by obtaining CT scan of these patients. In this study out of 26 patients 21 were fructose negative on seminal analysis and 5 had equivocal results. This study emphasized that absence of seminal fructose and low volume ejaculate in these patients may be due to agenesis of ejaculatory ducts rather than absence of seminal vesicles.⁵ However, in these two patients, seminal fluid examination had shown the presence of fructose indicating the presence of functioning seminal vesicles and patent ejaculatory ducts.

Conclusion

Isolated congenital absence of vas deferens can exist. Clinical examination is often unhelpful and misleading in these patients. Till there is sufficient development in scrotal imaging to localize the presence of vas, surgical exploration remains the only option for the diagnosis of these patients.

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