Unilateral segmental dysplasia of the vas deferens

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A healthy 35-year-old male presented for vasectomy after fathering two children. Due to difficulty palpating the left vas, the patient was taken to the operating room for scrotal exploration and vasectomy. The left vas was absent; however, a 1.2 cm pearly nodule was identified in the scrotum along its suspected course. This nodule was excised, found to contain thick white pasty fluid, and confirmed vas deferens by pathology. The patient was found to have normal kidneys on renal ultrasound and was indeed a carrier for cystic fibrosis gene mutations. We herein discuss management and implications of vasal anomalies.

Key Words: vasectomy, congenital absence of the vas deferens, infertility

Introduction

Every year in the United States anywhere from 175,000 to 354,000 men undergo vasectomy[1] and for various reasons, up to 6% of men who undergo vasectomy eventually pursue vasectomy reversal.[2] Prior to vasectomy, it is common practice for the surgeon to ensure the vas can be palpated bilaterally. Hunter first described congenital bilateral absence of the vas deferens (CBAVD) in 1737,[3] and since then unilateral cases of absent vas deferens have also been reported.[4] CBAVD accounts for about 6% of cases of obstructive infertility and 1%-2% of all male factor infertility.[5]

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Anger and Goldstein first characterized the unique finding of bilateral segmental dysplasia of the vas deferens (SDVD) in azoospermic men, as isolated segments of the vas deferens that are not connected to the epididymis or ejaculatory ducts.[6] They proposed that this finding, similar to that of CBAVD, may be related to CFTR gene mutations.[6] We describe the unique finding and management of a patient with unilateral SDVD.

Case report

An otherwise healthy 35-year-old male patient presented for vasectomy after fathering two children via intrauterine insemination for female factor infertility. He reported no prior history of genitourinary trauma, inguinal surgery, sexually transmitted infection or family history of infertility. On physical exam, the bilateral testes and right vas were palpably normal.

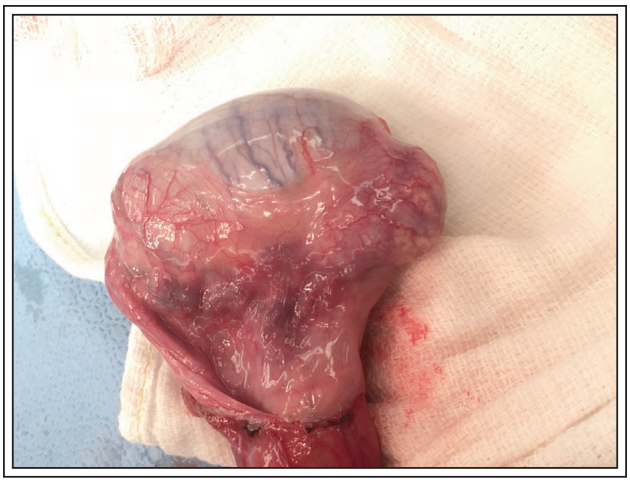

Figure 1. Testis with congested epididymis and absent vas deferens.

Due to difficulty palpating the left vas the patient was taken to the operating room for vasectomy with scrotal exploration.

Review of prior semen analysis demonstrated 1 mL total semen volume, 144.7 million sperm, 35% normal forms and 70% motility. Thus, right vasectomy was performed. On scrotal exploration the left epididymis appeared engorged and obstructed, Figure 1. The left vas appeared absent. A 1.2 cm pearly nodule was identified in the scrotum along the suspected course of the vas, not in communication with the epididymis or ejaculatory duct. This was excised, incised, and found to contain thick white pasty fluid similar to that seen in obstruction at time of vasectomy reversal, Figure 2. The segment was confirmed vas deferens on pathologic examination.

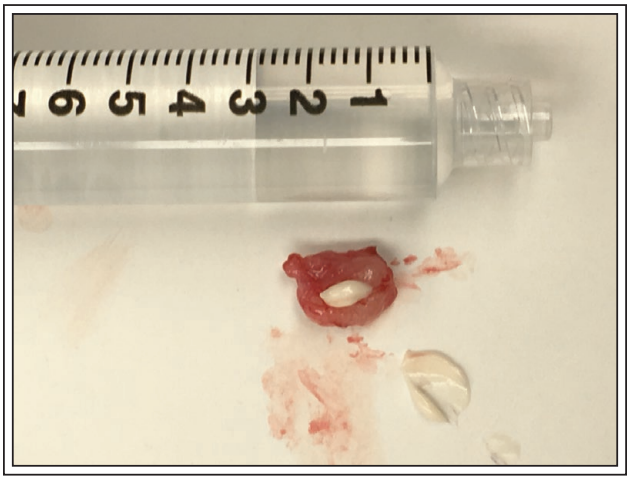

Figure 2. Vas segment with pearly white thick pasty fluid.

Discussion

To our knowledge, unilateral SDVD has not been previously described. In the previous report of bilateral SDVD all three men were being evaluated for obstructive azoospermia and were found to have isolated segments of the vas deferens 2 cm to 5 cm in length that were not connected to the epididymis or the ejaculatory ducts, with the gross appearance analogous to that of a string of sausages. The vasal fluid produced between two obstructed vas segments was thick, white, toothpaste-like material similar to that in men with secondary epididymal obstruction.[6] In our patient we identified a normal right vas and a solitary 1.2 cm short segment of vas in the scrotum not connected to the epididymis or the ejaculatory ducts on the left. Our patient's fertility was not impaired by unilateral disease, as evidenced by his previous semen parameters and successful childbearing.

Cases of absent or malformed vas deferens require unique management strategies, as failure of development of the mesonephric duct is thought to lead to absence or malformation of the vas deferens, and may potentially be associated with abnormalities of the epididymis, seminal vesicle, ureter and kidney.[7-8] In addition, numerable cystic fibrosis mutations have been associated with absent vas deferens, and reports describe a range of 72%-91% of men with CBAVD having at least one CFTR gene mutation.[9] Our patient had a normal renal ultrasound, but was indeed a CFTR mutation carrier. Testing for and proper counseling regarding CFTR genetic mutations is an important aspect of care for these patients.

Another interesting aspect of this study is regarding the origins of obstructed vasal fluid. At the time of vasectomy reversal all microsurgeons often evaluate both the gross and microscopic appearance of vasal fluid to determine the likelihood of obstruction. Thick, creamy fluid is often associated with secondary epididymal obstruction and this fluid rarely has any sperm or sperm parts present.[10] The origins of obstructed vasal fluid remain largely unknown and it is unclear if it may be derived from epididymis, vas deferens, or sperm. Based on our findings it is likely that the obstructive fluid is largely derived from the vas deferens itself, while it is difficult to draw any strong conclusions from this single interesting case.

Unilateral SDVD is a rare finding. Men with unilateral SDVD should also undergo upper tract imaging and testing for cystic fibrosis mutations. Current vasectomy guidelines do not to comment on surgical management of a man with unilateral vas deferens. The thick white pasty fluid produced in the obstructed segment of vas supports that similar fluid

seen in secondary epididymal obstruction is most likely derived from the vas deferens itself. As vasal fluid composition is affected by vasectomy and obstruction, further understanding of obstructive processes will help improve the management of obstructive infertility and congenital obstructive abnormities. □

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