Clinics in Surgery

Case Report
Published: 20 Jun, 2016

Epididymitis Due to Bilateral Ectopic Vas Deferens and Anorectal Malformation

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Abstract

Recurrent epididymitis and a history of anorectal malformation (ARM) may be secondary to an ectopic vas deferens to ureter described as persistent mesonephric duct syndrome. This is a case of a child with ARM found to have bilateral ectopic ureter to vas connection. The histopathology specimen here would suggest this is an ectopic ureter to vas. In a child with ARM that is undergoing nephrectomy, one should consider thorough exploration of the distal ureter to identify possible ectopic segments which may predispose him to epididymitis. This ectopia could explain the increased infertility rates of adults with a history of ARM.

Introduction

The acute scrotum, often secondary to testicular torsion, torsion of an appendix testis or epididymis, hernia, epididymo-orchitis, hydrocele, idiopathic scrotal edema, varicocele, and Henoch-Schonlein purpura, continues to be a diagnostic dilemma in emergency rooms as delayed or missed diagnosis can result in loss of gonadal tissue [1]. Epididymitis is a common cause of acute scrotum, though one that typically can be managed with medical therapy alone. Uncommon structural anomalies such as rectourethral fistula or ectopic vas deferens seen with persistent mesonephric duct syndrome, increases the risk of epididymitis and may warrant surgical correction. We present a case of recurrent bilateral epididymitis in a child with anorectal malformation (ARM) and discuss the embryological origin, associated defects, as well as management considerations.

Case Presentation

This is a case of a 3 year old male with history of high imperforate anus and bilateral high grade ureteral reflux disease identified at 2 days old. He was started on clean intermittent catheterization, nocturnal indwelling catheterization, and prophylactic antibiotics. He did well for two months, but then developed severe bilateral pyelonephritis with right sided renal abscess requiring left loop ureterostomy and right nephroureterectomy with preservation of the distal ureteral stump for possible future use as catheterizable stoma. He remained free of genitourinary infections. By 2 years of age, his imperforate anus was repaired, the ureterostomy was taken down, and he was found to have resolved left sided ureteral reflux on voiding cystourethrography (Figure 1). While waiting to undergo urodynamic testing, he developed four episodes of right sided epididymo-orchitis. He was subsequently taken for right ureteral stump excision and found to have an ectopic vas deferens exiting 2-3cm from the ureterovesical junction. Six months later, following an episode of now left epididymo-orchitis, his video urodynamics demonstrated a left sided ectopic vas deferens to the distal ureter and recurrence of his vesicoureteroreflux (Figure 2). He was again taken to the operating room and underwent left ureteroscopy and placement of a ureteral catheter into this ectopic segment, followed by a left inguinal exploration where the ureteral catheter was isolated and the ectopic segment was ligated and excised. Since, he has been free of any recurrent epididymo-orchitis episodes.

Discussion

Epididymitis is associated with anorectal malformation in 1.2-6.1% [2]. This uncommon urologic condition is more often associated with existing rectourethral fistula leading to chronic urinary tract infections or anatomic alterations due to fistula insertion into the posterior urethra [3]. Despite repair of the fistula tract, episodes of epididymitis may recur in more than one third of cases. Ectopic vas deferens is a rare cause of epididymitis though is often associated with anorectal malformation. Upwards of 30 cases of vasa ectopia have been described since 1895, with only 6

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Received Date: *22 May 2016*
Accepted Date: *15 Jun 2016*
Published Date: *20 Jun 2016*

Citation:
Upadhyay J, Allemang T. Epididymitis Due to Bilateral Ectopic Vas Deferens and Anorectal Malformation. Clin Surg. 2016; 1: 1034.

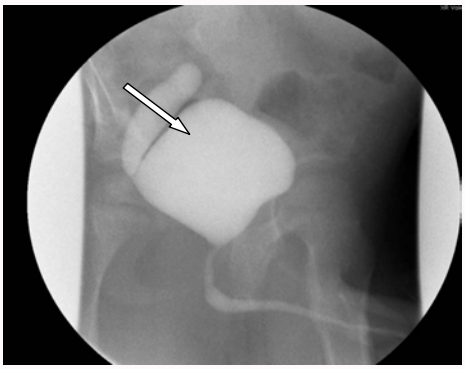

Figure 1: Initial voiding cystourethrogram without evidence of bilateral ectopic vas deferens. Arrow shows reflux into right ureter.

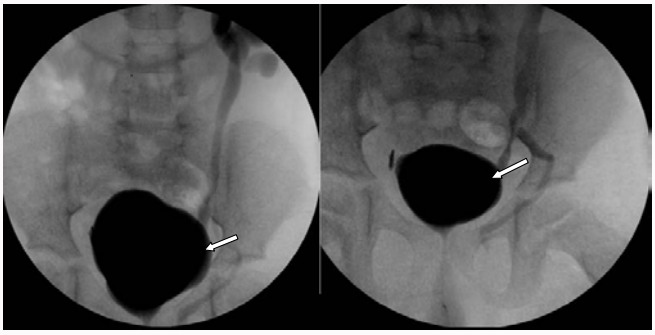

Figure 2: Left sided ectopic vas deferens (arrow) to left ureter on voiding cystourethrography.

previously described with bilateral ectopia [4-6]. In addition to the association with anorectal malformation, this is frequently associated with ipsilateral renal anomalies to include dysplasia and agenesis, as well as ipsilateral ureteral obstruction or reflux [5].

Ectopic vas deferens has been attributed to persistent mesonephric duct syndrome (PMDS), which is defined by a common distal ureteral segment, or common mesonephric duct, that drains both a proximal ureter, as well as an ectopic vas deferens [4,6]. This common excretory duct may be connected along any length of the collecting system, from the renal pelvis to the bladder [5]. The embryological origin of PMDS and its association with ARM is likely secondary to its temporally associated maldevelopment. As the ureteric bud grows cranially, the common mesonephric duct is resorbed caudally into the cloaca to form the trigone. Meanwhile, the cloaca begins dividing during the fourth week of gestation into the anterior urogenital sinus and the posterior rectum. The now resorbed common mesonephric duct has two separate openings, the distal ureter into the trigone, and the vas deferens as it migrates and fuses with the posterior urethra [4,5,7]. It is the incomplete fusion of the common mesonephric duct with the cloaca which is believed to result in the ectopic location of the vas deferens to the urinary collecting system.

As was described by Schwartz and Stephens et al. [6] the ectopic vasal segment seen in PMDS histologically resembles ureteral tissue, which was found in this case as seen in Figure 3.

Histological cross sections of the ectopic vas segment appears to have an transitional cell epithelial lining as would be expected with ureteral tissue, rather than the pseudostratified epithelium seen with normal vas deferens. Additionally, the three muscle layers of an inner longitudinal, middle circular and outer longitudinal muscle fibers

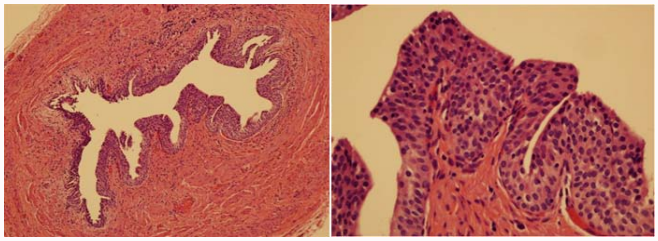

Figure 3: (Left) 10x Microscope view showing Ectopic Ureter/Vas, (Right) 40x microscope view showing transitional cell epithelium similar confirming a ureteral histologic appearance of ectopic vas segment.

typically seen in the normal vas deferens, is absent in the ectopic vas segment. This histological finding may represent a heteroplasia that occurs due to abnormal signaling during the differentiation of the mesonephric duct into the vas deferens. As such, it is arguable whether this truly represents an ectopic ureter to vas connection as is seen histologically here and in previous published cases [6,7], versus an ectopic vas to ureter connection which is described embryologically by Vordermark and Schwartz [4,6]. Another possibility is that the urothelial appearance of this ectopic vas may be secondary to a metaplasia which develops due to the continued exposure of the vasal tissue to refluxing urine either perinatal or postnatal.

With the reflux of urine through the common excretory duct and into the ectopic vas deferens, acute and recurrent epididymitis may occur. The chronic inflammation within the ejaculatory system and testis, leads to irreversible scarring and may account for the 20% of associated infertility in boys with anorectal malformation [8]. While attempts have been made to endoscopically treat this reflux of urine by the injection of UROCOL into the common distal excretory segment to reduce UTI, epididymitis and infertility, long term results of such treatment is unknown [9]. In the largest case series of epididymitis associated with anorectal malformation, a therapeutic algorithm was proposed. In a child with recurrent unilateral epididymitis and good bilateral renal function, one could consider ipsilateral ureteral reimplant to allow for the investigation of an ectopic vas segment into either the bladder or ureter, or to reduce reflux urine into a potential common mesonephric duct segment. If a child has poor ipsilateral renal function, then a nephroureterovasectomy should be performed. This same algorithm attempts to address the situation of bilateral epididymitis though attributes this to a more distal urinary abnormality such as neurovesical dysfunction, urethra-ejaculatory duct reflux, urethral stricture, or valves (Raveenthiran) [2].

Although seemingly rare, our case of bilateral persistent mesonephric duct syndrome causing ectopic vaso-ureter connection, eventually had successful treatment with bilateral vasectomies as would have been recommended by the previously described algorithm. There are some technical lessons to be gained from this case. First, occult ectopic vas-ureter connection as was found on the left side in this case may be more prevalent than previously believed, thus contributing to the 20% infertility rate in boys with ARM. Second, applying the previously described algorithm to any boy with ARM and dysplastic kidney requiring nephrectomy, even in the absence of epididymitis, one should consider complete ureterectomy with concomitant exploration for possible ectopic vas-ureter segment. Third, a voiding cystourethrogram that does not show reflux into an ectopic segment does not rule out the absence of an ectopic vas. High clinical suspicion is needed in cases of recurrent epididymitis. Finally, in the setting of bilateral epididymitis and ARM, evaluation should

not only be limited to cystoscopy, but also include bilateral retrograde urethrograms looking for occult vasal ectopia.

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