Abdom Imaging 18:193–195 (1993)

Abdominal Imaging

Duplicated Mullerian Duct Remnants Associated with Unilateral Renal Agenesis

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Abstract. A case of duplicated Mullerian duct remnants associated with unilateral renal agenesis in a male patient is described. This association is not uncommon in females, but is rarely recognized in male patients. The possibility of Mullerian duct cyst or enlarged prostatic utricle should be considered in the differential diagnosis of a pelvic cyst in a male patient with unilateral renal agenesis.

Key words: Mullerian duct cyst—Enlarged prostatic utricle—Renal agenesis.

Mullerian duct cyst and enlarged prostatic utricle are rare congenital anomalies of promesonephric (Mullerian duct) origin. They may be asymptomatic or present with symptoms of infection or obstruction. There is an association with genital anomalies and intersex states, and a rare association with renal agenesis. We describe a case with both Mullerian duct cyst and enlarged prostatic utricle in a patient with unilateral renal agenesis. It was investigated with intravenous urography, micturating cystourethrography, isotope renography, and ultrasound, and there is surgical correlation. Only one previous case of duplicated Mullerian duct remnants and unilateral renal agenesis in a male patient has been described [1].

Case Report

A 7-year-old boy was referred with recurring epididymoorchitis. General physical examination was normal. Urine culture repeatedly showed a significant growth of *Escherichia coli*. Subsequent chromosome studies revealed a normal male karyotype.

Ultrasound showed a cystic mass behind the bladder with a smaller cystic mass inferior to this (Fig. 1). The left kidney was large and no right kidney was demonstrated. An intravenous urogram showed a solitary large left kidney (Fig. 2). A micturating cystourethrogram showed a large volume bladder which emptied completely on micturition, with no evidence of reflux. During micturition a cystic space filled from the posterior urethra (Fig. 3). Cystoscopy and retrograde urethral catheterization were performed. A sinus track was identified leading posteriorly from the verumontanum. Contrast injection confirmed this to communicate with the previously identified cystic cavity (Fig. 4) and a diagnosis of an enlarged prostatic utricle was made.

An isotope renogram showed good renal function on the left with free drainage. No functioning renal tissue was identified on the right.

At surgery the bladder was exposed and opened vertically. An incision was made through the posterior wall of the bladder from the trigone to the dome. The enlarged prostatic utricle was identified and dissected from the surrounding tissues. During dissection a separate cavity with no clear connection to the urethra was identified inferior to the utricle. Both the enlarged prostatic utricle and the second cyst were excised. The vas deferens were identified entering the utricle and cyst. The right was reimplanted into the bladder and the left was ligated due to lack of length.

Histology showed both utricle and cyst to be similar. Both had a muscularis mucosa resembling that of the bladder and were lined by transitional epithelium in places. Two ducts lined by pseudostratified columnar epithelium were present and were thought to represent either vas deferens or seminal vesicle. There was moderately severe inflammatory cell infiltrate in the cyst walls. The site and histology of the cyst were thought to be compatible with a diagnosis of a Mullerian duct cyst.

Discussion

Renal agenesis is caused by failure of development of the metanephros in utero. The metanephros or permanent kidney arises from the base of the mesonephros between the 5th and 8th weeks of embryogenesis. The remaining mesonephros gives rise to the Wolffian duct laterally and the Mullerian duct medially. In the male, the Wolffian duct differentiates into the epididymis, vas deferens, ejaculatory duct, and seminal vesicles, whereas the Mullerian duct

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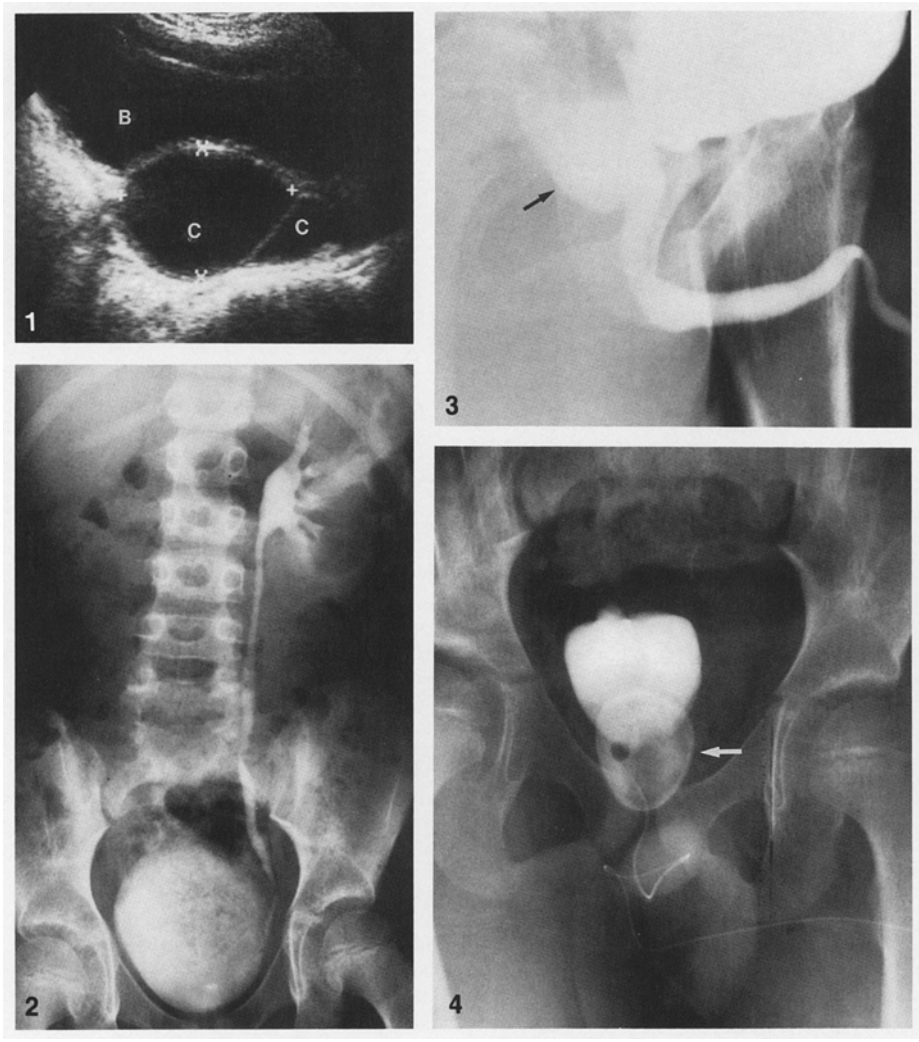

Fig. 1. Sagittal view from a sonogram of the pelvis showing two cystic lesions (*C*) lying posterior to the bladder (*B*).

Fig. 2. A 15-min film from an excretory urogram showing a solitary large left kidney.

Fig. 3. Micturating cystourethrography showing an enlarged prostatic utricle (*arrow*) filling from the posterior urethra.

Fig. 4. Retrograde urethrogram demonstrating the enlarged prostatic utricle (*arrow*).

involutes to become the appendix of the testes and the prostatic utricle. Abnormal evolution of the mesonephros during this period therefore has the potential to affect all these structures. Male genital tract malformations have been described in 20–70% of cases of renal agenesis [2]. The most common abnormalities are agenesis or cyst of the seminal vesicles [2]. Mullerian duct malformations, such as Mullerian duct cyst or enlarged prostatic utricle, are less common. A literature review in 1978 found only seven cases of renal agenesis associated with enlarged prostatic utricle or Mullerian duct cyst [3]. Since then four further cases have been described [1, 3–5].

Some authors do not distinguish between enlarged prostatic utricle and Mullerian duct cyst [3]. Despite a similar embryological etiology there are structural and clinical differences which have recently been emphasized [6]. An enlarged prostatic utricle usually presents in the first or second decade. It is commonly associated with hypospadias or intersex states. It usually communicates with the posterior urethra and does not extend beyond the prostate. A Mullerian duct cyst presents later, often in the third or fourth decade. It does not communicate with the prostatic urethra and may grow to a considerable size [4, 5]. It is usually an isolated abnormality which may explain its later presentation. Presentation is usually due to obstructive symptoms, urinary tract infection, or epididymitis [3]. The vas deferens frequently enters the cyst, thus explaining the increased frequency of epididymitis which can occur with an enlarged prostatic utricle or Mullerian duct cyst [3].

The presence of both a Mullerian duct cyst and an enlarged prostatic utricle in a patient with renal agenesis is very rare. Only one other case has been described [1]. Normally, the paired Mullerian ducts fuse in the midline during the 8th week in utero. Failure to do so can give rise to the anomalies described in this case. Duplicated Mullerian duct remnants associated with renal agenesis have been described more often in women because of the frequent association with unilateral hematocolpos [7]. The rarity of cases such as ours in the literature is therefore probably due to underdiagnosis.

Most cases in the literature have been investigated with voiding cystourethrography and/or retrograde urethrography. These tests will not detect Mullerian duct cysts because of their lack of communication with the posterior urethra. In the past decade the advantages of ultrasound [2], computed tomography [4], and magnetic resonance [8] have been described. Ultrasound in particular has the advantage of rapid visualization of the whole genitourinary system [2].

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