ABSTRACT

Mullerian duct cyst is an uncommon congenital anomaly. It is usually small, asymptomatic, midline, cystic lesion, located behind the superior half of the prostatic urethra and connected to the verumontanum by a thin stalk. Rarely a mullerian duct cyst may be associated with renal agenesis and hypospadias. A case of mullerian duct cyst with unilateral renal agenesis and hypospadias is being reported.

Keywords: Mullerian duct cyst; prostate; Ultrasound, CT Scan, Magnetic Resonance Imaging

Case Report

A 5-year old male child presented with irritative lower urinary tract symptoms and hypospadias. On abdominal ultrasonography, there is a 15x6x3.8 cm unilocular cystic nature mass lesion seen in the pelvis. It was located posterior to bladder base, extending superiorly to the level of the iliac vessels and compressing the bladder anteriorly. Right kidney could not be visualized at the normal localization.

Contrast enhanced computerized tomography (CT) abdomen and pelvis confirmed the presence of cystic lesion posterior to the urinary bladder in pelvis and had no communication with urinary system. Agenesis of right kidney was seen.

A pelvic MRI showed that lesion was hyperintense on STIR and T2 weighted images and hypointense on T1 weighted images. It was located high above the base of the prostate. The seminal vesicles are displaced superiorly by this cystic mass lesion. There was no ectopic ureteral orifice or dilatation of the ejaculatory duct. The case was diagnosed as mullerian duct cyst with unilateral renal agenesis and hypospadias. The patient has minimal symptoms, hence surgical resection was not considered.

Discussion

Mullerian duct cyst is a remnant of the caudal ends of the fused mullerian ducts which typically regresses in utero. These cysts are typically located in the midline, posterior to the bladder, originating in the region of the verumontanum. Mullerian duct cysts do not communicate with the posterior urethra nor contains any sperm. It was connected to the verumontanum by a thin stalk. Patients are usually asymptomatic but may present with urinary retention, urinary tract infection, or symptoms of ejaculatory duct obstruction such as hemospermia. The peak clinical incidence of mullerian duct cysts is in the age range of 20-40 years. Few cases are reported in infancy. Mullerian duct cyst is not usually associated with other congenital abnormalities of the urinary tract but occurs in isolation. Rarely, a Mullerain duct cyst may be associated with renal agenesis. However this case had unilateral renal agenesis with hypospadias.

Differentials are prostatic utricle cyst, ejaculatory duct cyst, cyst of the vas deferens, seminal vesicle cysts, ectopic ureteroceles and abscesses. Mullerian duct and Prostatic utricle cysts are in midline location. Ejaculatory duct cyst, cyst of the
vas deferens, seminal vesicle cysts, and ectopic ureteroceles are cystic lesions seen in the paramedian location. If these cysts reach big in sizes they displace the bladder anteriorly and the colon posteriorly and cause symptoms.6 (Tab. 1)

<table>
<thead>
<tr>
<th>Age of presentation</th>
<th>Mullerian duct cysts</th>
<th>Prostatic utricular cysts</th>
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<tbody>
<tr>
<td>Third to fourth decade</td>
<td>Round</td>
<td>Tubular</td>
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<tr>
<td>Large</td>
<td>Remnants of the mullerian duct</td>
<td>Uncommon but connected to the verumontanum by a stalk</td>
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<tr>
<td>Uncommon but connected to the verumontanum by a stalk</td>
<td>Common</td>
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Table 1: Mullerian duct cyst vs prostatic utricular cyst

Most utricular cysts are diagnosed in childhood because of association with hypospadias, pseudohermaphroditism, and cryptorchidism. Utricular cyst rises from the verumontanum and communicates with the posterior urethra, a feature that helps distinguish it from a mullerian duct cyst radiologically. Mullerian duct cysts are connected to the verumontanum by a stalk but do not communicate with the posterior urethra. They extend above the prostate if large. Carcinoma is a rare potential complication.2,7 Sometimes mullerian duct cyst and prostatic utricle cyst are difficult to be differentiated on imaging basis. Differentiation is of academic importance. There is no evidence that mullerian duct cyst originate from the Müllerian duct remnant, at least in the epithelial lining.8

In this case transabdominal sonogram demonstrated the cystic mass clearly, but its origin could not be defined (Fig. 1A). Sonography, in particular transrectal sonography, is an excellent tool for the evaluation of Mullerian duct cyst. In this case, patient had minimal symptoms so transrectal ultrasonography and surgical resection was not performed. Post contrast CT pelvis reveals contrast filled urinary bladder and nonenhancing cystic lesion with no communication with bladder/urethra as confirmed on delayed scan (Fig 1B and 3A).

On MRI, the lesion was hyperintense on STIR and T2 weighted MR images and hypointense on T1 weighted images (Fig. 2A, 2B and 2C). The cyst did not show any contrast enhancement. The seminal vesicles were displaced by the cystic mass lesion. MRI has been reported to be useful in the diagnosis of Mullerian duct cyst by showing signal characteristic of the mucinous or hemorrhagic cystic component.9 However, it may show increased T1-weighted and T2-weighted signal intensity reflecting increased concentration of mucinous material or hemorrhage. Surgical excision of a mullerian duct cyst depends on the size and location of the cyst and the presence of clinical symptoms. Almost 60% of adults diagnosed with a mullerian duct cyst do not experience any cyst-related symptoms or ejaculatory-fertility impairment, so treatment only recommended in symptomatic or infertile patients.10

In conclusion, mullerian duct cyst associated with unilateral renal agenesis and hypospadias is a rare congenital anomaly. MRI accurately defines ana-
Anatomic relationship when one is planning to excise a mullerian duct cyst due to multiplanar imaging capacity, superior soft tissue contrast, and absence of ionizing radiation.

Figure 2: Pear shaped cystic nature mass lesion seen hyperintense on STIR and T2 weighted images (A,B) and hypointense on T1 weighted images (C).

Figure 3: Sagittal reformatted CT images (A) and T2W MR images (B) show that there is no communication between cystic lesion and urinary bladder/urethra.
References


