


Zinner's Syndrome: Case report of a Developmental Anomaly of the Mesonephric Duct

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ABSTRACT

Zinner's syndrome is a rare congenital malformation characterized by the association of unilateral renal agenesis with ipsilateral seminal vesicle cyst and ejaculatory duct obstruction. Most patients are asymptomatic until the third or fourth decade of life when the syndrome is associated with dysuria, perineal pain, infertility, and painful ejaculation.

In this report, we present the common imaging findings of this rare developmental anomaly involving the mesonephric duct in a 48-year-old male patient experiencing pelvic pain, recurrent dysuria, and pollakiuria.

CASE REPORT

CASE REPORT

A 48-year-old adult male presented to the urology department of our Hospital with pelvic pain and pollakiuria. The patient also reported previous, similar episodes with an increased frequency of urination and dysuria. There was no history of lower urinary tract symptoms, painful ejaculation, hematuria, or trauma. Past medical history was negative except for the presence of mild hypertension. Physical examination was unremarkable. The patient had a recent CT examination in another Institute, performed without the administration of contrast media, for suspected calculosis in which was reported the absence of the left kidney and the presence of a non-specific mass in the pelvic area (Figure 1). For further evaluation, the patient was referred to our Radiology department. An ultrasound revealed the absence of the left kidney with hypertrophied right kidney and a well-defined anechoic structure displacing the bladder anteriorly (Figure 2).

To confirm the origin of the cystic lesion, an MRI examination was performed. MRI confirmed the absence of the left kidney and showed a large cystic lesion replacing the left seminal vesicle of approximately 60x43x43 mm (Figure 3A). The seminal vesicle cyst was mildly hyperintense on T2 and T1-weighted images, due to hemorrhage or proteinaceous content and showed no restriction or enhancement after administration of contrast media (Figure 3B-3C-3D and Figure 4A-4B). Additionally, a superior blind-ending tubular structure was also apparent, running along the left iliac vessels and draining into the ipsilateral seminal vesicle, consistent with an ectopic atretic ureter (Figure 5). Based on these findings, a diagnosis of a seminal vesicle cyst with left renal agenesis was suggested and the patient was referred to a urologist.

DISCUSSION

BACKGROUND

Zinner's syndrome is associated with an embryologic abnormality that develops in the distal portion of the mesonephric or Wolffian duct between the 4th and 13th gestational week. The incomplete migration of the ureteric bud is responsible for the lack of differentiation of the metanephric blastema leading to ipsilateral renal agenesis and atresia of the ejaculatory duct [1].

First described by Zinner in 1914, this congenital abnormality had been reported in about 200 cases as of 2020 [2] and is considered to be the male equivalent of the Mayer-Rokitansky-Küster-Hauser syndrome [3]. The diagnosis is usually made between the 3rd and the 4th decades of life due to enlargement of the seminal vesicle cysts. By this age, cysts can grow larger than 6 cm and become symptomatic, with the most common symptoms including abdominal, perineal, and pelvic pain. Other possible symptoms may be ejaculatory pain, dysuria, hematuria, urinary tract infections, and symptoms of epididymitis and prostatitis. Differential diagnosis involves several cystic diseases of other pelvic organs, including prostatic utricle cysts, ejaculatory duct cysts, prostatic cysts, diverticula of the ampulla of vas deferens, ureterocele, tumors and abscesses [4].

Cyst location, along with other developmental abnormalities such as renal agenesis or anomalies of the external genitalia, can aid in differential diagnosis. Congenital anomalies of the kidneys and urinary tracts (CAKUT) are embryonic disorders that arise during development and result in a spectrum of defects in the kidneys and outflow tracts which include the ureters, the bladder, and the urethra with an estimated prevalence of 4–60 per 10,000 births [5].

IMAGING FINDINGS

Radiological imaging plays an essential role in the evaluation of such cystic lesions. [1] Ultrasonography, often the initial investigative technique, is capable of detecting cystic masses as anechoic thick-walled cysts. Possible internal echoes may be found in the case of infection or hemorrhage.

CT and MRI scans allow for panoramic evaluation of the pelvis, with the MRI being the modality of choice for the accurate anatomical demonstration of the male genital tract [6].

CT is usually reserved for further evaluation when MRI is unavailable or contraindicated, but the acquisition with contrast media is mandatory to avoid misinterpretation.

In MRI, the seminal vesicle cyst appears as a fluid-filled image with hypointensity on T1 weighted sequences, strong hyperintensity on T2, and the absence of areas of restriction on DWI or enhancement after the administration of a contrast agent. Exceptions may occur in cases with high protein content, which can reveal an increased T1-weighted signal and possible mild restriction on DWI, with the absence of enhancement. The demonstration of communication between the cystic lesion and the seminal vesicle confirms the origin of the cyst.

Treatment

In cases where symptoms such as recurrent urinary tract infections, hematospermia, or lower abdominal pain are present, treatment may be necessary.

Antibiotics may be prescribed to treat and prevent urinary tract infections, which are common in individuals with Zinner's syndrome. Surgical removal of seminal vesicle cyst is suggested in cases where the seminal vesicle cyst is larger than 6 cm, the approach may involve open surgery or minimally invasive techniques such as laparoscopy [7].

TEACHING POINTS:

Radiological imaging is essential in the diagnosis of Zinner's syndrome. Ultrasound could be the first imaging technique of investigation. MRI is the gold standard technique for the evaluation of the urogenital system. A CT could be performed if MRI is unavailable, but the use of contrast medium is mandatory.

AUTHORS' CONTRIBUTIONS

Mirco Cleva: corresponding author who wrote the case report

Luca Montaldo: performed the US examination

Giovanna Graziani: performed the MRI examination

Ennio Bruschi and Massimo Valentino: responsible for the correction of the writing

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DISCLOSURES

I have no disclosures to report.

CONSENT

Did the author obtain written informed consent from the patient for submission of this manuscript for publication? Yes

KEYWORDS

Zinner's syndrome, MRI, US, TC, Wolffian duct abnormality, Renal agenesis, Seminal vesicle cyst,

1) What are the main features of Zinner's syndrome? (More than one answer is correct)

- Renal Agenesis (**applies**)
- Ectopic Kidney
- Seminal vesicle cyst (**applies**)
- Ejaculatory duct obstruction (**applies**)
- Duplex Kidney

1- Renal Agenesis. (**applies**) [Zinner's syndrome is a rare congenital malformation characterized by the association of unilateral renal agenesis with ipsilateral seminal vesicle cyst and ejaculatory duct obstruction]

2- The congenital anomaly of the kidneys associated with Zinner's syndrome is the absence of the homolateral Kidney. [The incomplete migration of the ureteric bud is responsible for the lack of differentiation of the metanephric blastema leading to ipsilateral renal agenesis and atresia of the ejaculatory duct]

3- Seminal vesicle cyst. (**applies**) [Zinner's syndrome is a rare congenital malformation characterized by the association of unilateral renal agenesis with ipsilateral seminal vesicle cyst and ejaculatory duct obstruction]

4- Ejaculatory duct obstruction. (**applies**) [Zinner's syndrome is a rare congenital malformation characterized by the association of unilateral renal agenesis with ipsilateral seminal vesicle cyst and ejaculatory duct obstruction]

5- The congenital anomaly of the kidneys associated with Zinner's syndrome is the absence of the homolateral Kidney. [Cyst location, along with other developmental abnormalities such as renal agenesis or anomalies of the external genitalia, can aid in differential diagnosis]

2) Which demographic is most affected by Zinner's syndrome?

- Male population aged between 1-20 years old
- Female population aged between 1-20 years old
- Male population aged between 30-40 years old (**applies**)
- Female population aged between 30-40 years old
- Male population aged over 50 years old

1- Seminal vesicle cysts tend to enlarge from the age of 20 to reach their maximum dimensions around the third and fourth decades. [The diagnosis is usually made between the 3rd and the 4th decades of life due to enlargement of the seminal vesicle cysts]

2- The syndrome affects the male population. [Zinner's syndrome is considered to be the male equivalent of the Mayer-Rokitansky-Küster-Hauser syndrome]

3- Male population aged between 30-40 years old. **(applies)** [At this age cysts can be larger than 6 cm and symptomatic, with the most common symptoms being abdominal or perineal, and pelvic pain]

4- The syndrome affects the male population. [Zinner's syndrome is considered to be the male equivalent of the Mayer-Rokitansky-Küster-Hauser syndrome]

5- After the age of 40 the cysts tend to decrease in size. [The diagnosis is usually made between the 3rd and the 4th decades of life due to enlargement of the seminal vesicle cysts]

3) Which radiological approach is most appropriate in the evaluation of Zinner's syndrome?

- First level MRI examination
- Ultrasound as first level examination followed either by MRI or non-contrast tomography
- Single ultrasound examination
- Ultrasound as first level examination followed either by MRI or contrast-enhanced tomography to make a differential diagnosis **(applies)**
- Contrast-enhanced ultrasound (CEUS)

1- MRI is the gold standard for the evaluation of the urogenital system but is usually a secondary-level examination. [Ultrasonography is often the initial investigation technique able to detect a cystic mass]

2- It is the administration of the contrast medium that allows the differential diagnosis of cystic lesions of the pelvis. [CT is usually reserved for further evaluation when MRI is unavailable or contraindicated but is mandatory the acquisition with contrast media to avoid misinterpretation]

3- The ultrasound can detect but not perform a differential diagnosis between pelvic lesions. [Ultrasound could be the first imaging technique of investigation. MRI is the gold standard technique for the evaluation of the urogenital system]

4- Ultrasound as first level examination followed either by MRI or Contrast tomography to make a differential diagnosis. **(applies)** [Ultrasonography is often the initial investigation technique able to detect a cystic mass...CT and MRI scans allow for panoramic evaluation of the pelvis, with the MRI being the modality of choice for the accurate anatomical demonstration of the male genital tract.]

5- The administration of gas-filled microbubbles helps to better visualize organs and blood vessels within the abdomen and pelvis, however, the modality lacks the panoramic view of tomography and MRI examinations to assess the urogenital system. [CT and MRI scans allow for panoramic evaluation of the pelvis, with the MRI being the modality of choice for the accurate anatomical demonstration of the male genital tract.]

4) Which imaging is the best for anatomical evaluation of the urogenital system?

- Tomography with administration of contrast medium
- Non-contrast tomography
- Ultrasound
- MRI **(applies)**
- Positron emission tomography (PET)

1- MRI is the gold standard for evaluation of the urogenital system. [CT is usually reserved for further evaluation when MRI is unavailable or contraindicated]

2- The contrast media agent is mandatory for optimal visualization of the urogenital system. [A CT could be performed if MRI is unavailable, but the use of contrast medium is mandatory]

3- Ultrasound lacks the panoramic view of tomography and MRI examinations. [Ultrasound could be the first imaging technique of investigation. MRI is the gold standard technique for the evaluation of the urogenital system.]

4- MRI. **(applies)** [MRI is the gold standard technique for the evaluation of the urogenital system]

5- Positron emission tomography is not indicated for an anatomic evaluation. [MRI is the gold standard technique for the evaluation of the urogenital system]

5) Which are the main features of a seminal vesicle cyst at MRI? (More than one answer is correct)

- T1-sequence hypointensity and T2-sequence hyperintensity with the absence of Diffusion Weighted Imaging restriction or enhancement in cysts with fluid content **(applies)**
- Mild hyperintensity on T1-weighted and T2-weighted sequences with possible mild restriction on Diffusion Weighted Imaging with the absence of contrast enhancement if presence of hemorrhage or proteinaceous content within the cyst **(applies)**
- Mild hyperintensity on T1-weighted and T2-weighted sequences with possible mild restriction on Diffusion Weighted Imaging with strong enhancement after contrast media administration if presence of hemorrhage or proteinaceous content within the cyst
- Mild hyperintensity on T1-weighted and T2-weighted sequences with possible mild restriction on Diffusion Weighted Imaging in cysts with fluid content
- Mild hypointensity on T1-weighted and T2-weighted sequences with the absence of Diffusion Weighted Imaging restriction or enhancement after contrast media administration in cysts with fluid content

1- Liquids on MRI are hypointense on T1-weighted sequences and hyperintense in T2-weighted sequences without restriction or abnormal enhancement after contrast media administration. [At MRI, the seminal vesicle cyst appears as a fluid image with hypointensity on T1 weighted sequences and a strong hyperintensity on T2 and absence of areas of restriction on DWI or enhancement after administration of contrast media agent]

2- Hemorrhagic or proteinaceous content shows mild hyperintensity on both T1-weighted and T2-weighted sequences with possible restriction on diffusion. [possible exceptions are cases of high protein content that reveal an increased T1-weighted signal and possible mild restriction on DWI.]

3- Complicated cysts with blood or proteinaceous content do not show enhancement after contrast media administration. [possible exceptions are cases of high protein content that reveal an increased T1-weighted signal and possible mild restriction on DWI with the absence of enhancement.]

4- Mild Hyperintensity on T1-weighted and T2-weighted images with possible mild restriction on Diffusion Weighted Imaging are characteristics of complicated cysts. [At MRI, the seminal vesicle cyst appears as a fluid image with hypointensity on T1 weighted sequences and a strong hyperintensity on T2 and absence of areas of restriction on DWI or enhancement after administration of contrast media agent]

5- Cysts with fluid content are hypointense on T1-weighted sequences and hyperintense on T2-weighted sequences. [At MRI, the seminal vesicle cyst appears as a fluid image with hypointensity on T1 weighted sequences and a strong hyperintensity on T2]

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FIGURES



Figure 1: 48-year-old male with pelvic pain and dysuria.

Findings: Non-specific mass (*white arrow*) with intermediate density (mean value of 38 H.U.). The mass lies posteriorly to the bladder (*asterisk*) at the same plane of the right seminal vesicle (*black arrowhead*).

Technique: CT abdomen without contrast. Axial view 2.5mm slice thickness.

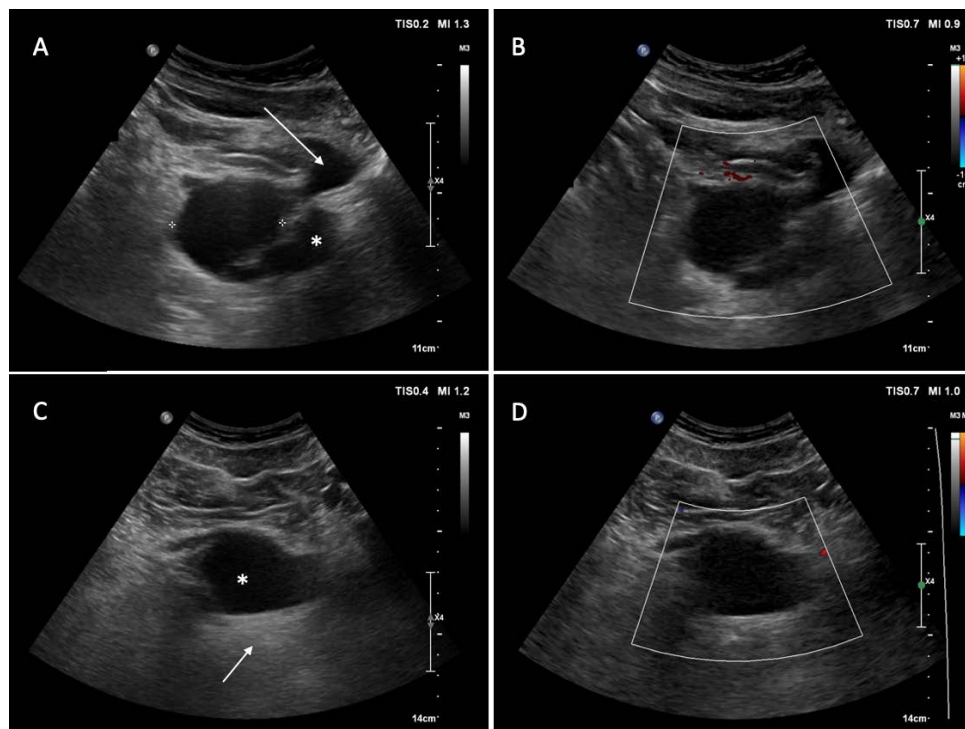


Figure 2: 48-year-old male with pelvic pain and dysuria.

Findings: Sagittal B-mode ultrasound on the median plane, the cystic lesion (*between calipers*) is posterior to the bladder (*arrow*) and superior to the prostate in continuity with dilated seminal vesicles (*asterisk*) (2A); Axial view of the cystic lesion (*asterisk*), that shows smooth margins, a fluid content and posterior acoustic enhancement (*arrow*) (2C); B-mode ultrasound with color-Doppler on the in the same plane as A and C that demonstrate no vascularization of the cystic lesion (2B, 2D).

Technique: Gray scale (A and C) and Color-Doppler (B and D) ultrasound imaging of the pelvis using 1-5 MHz convex probe. A and B) Sagittal view, C and D) Axial view.

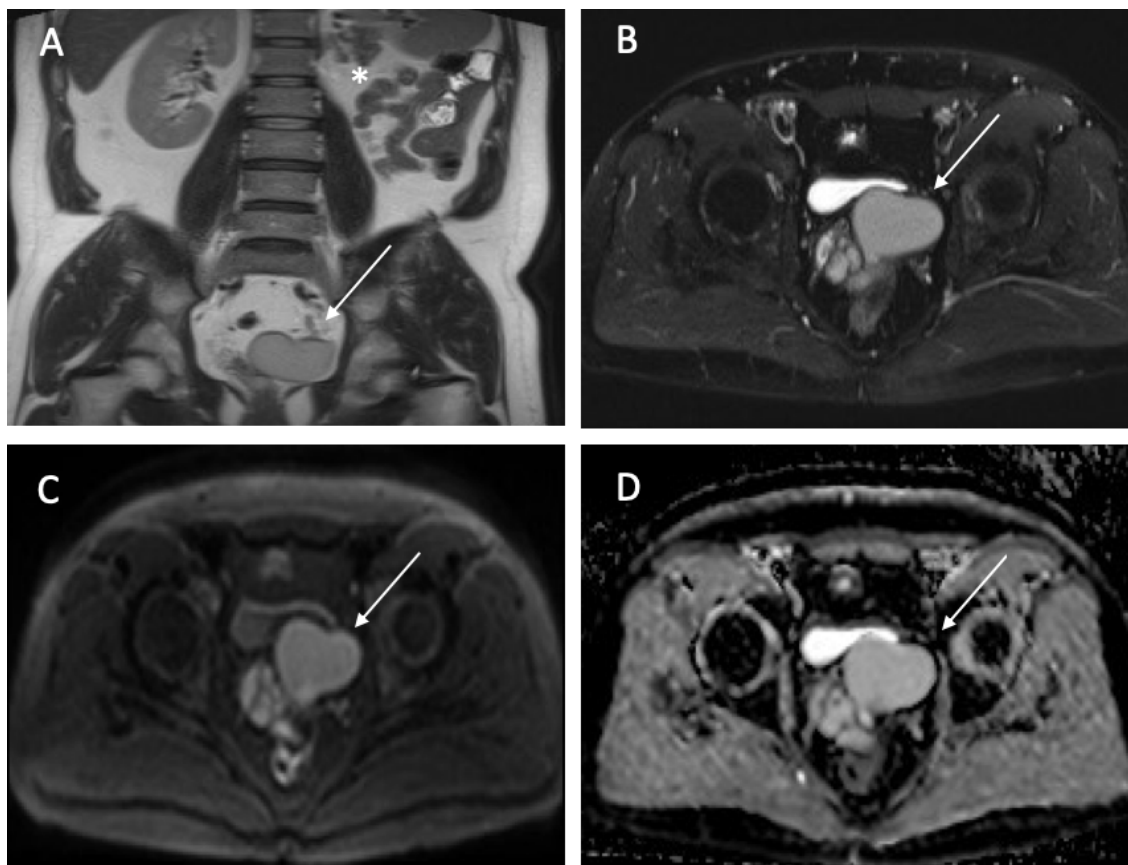


Figure 3: 48-year-old male with pelvic pain and dysuria.

Findings: Coronal non-contrast T2-weighted MR image confirms the presence of a large cystic lesion mildly hyperintense in the pelvic area (*arrow*) with the absence of the homolateral Kidney (*asterisk*)(3A); Axial non-contrast T2 HASTE with fat saturation underline the mild hyperintensity of the left seminal vesicle cyst (*arrow*)(3B); DWI and ADC map show no restriction of the cyst (3C,3D).

Technique: MRI 1.5 Tesla. 16 mL Gadoteridol (Pro-Hance) MR imaging of the pelvis. A) Coronal non-contrast T2-weighted MR image, B) Axial non-contrast T2 HASTE with fat saturation. C and D) Axial high b-value DWI sequence ($b = 800 \text{ s/mm}^2$) and ADC map.

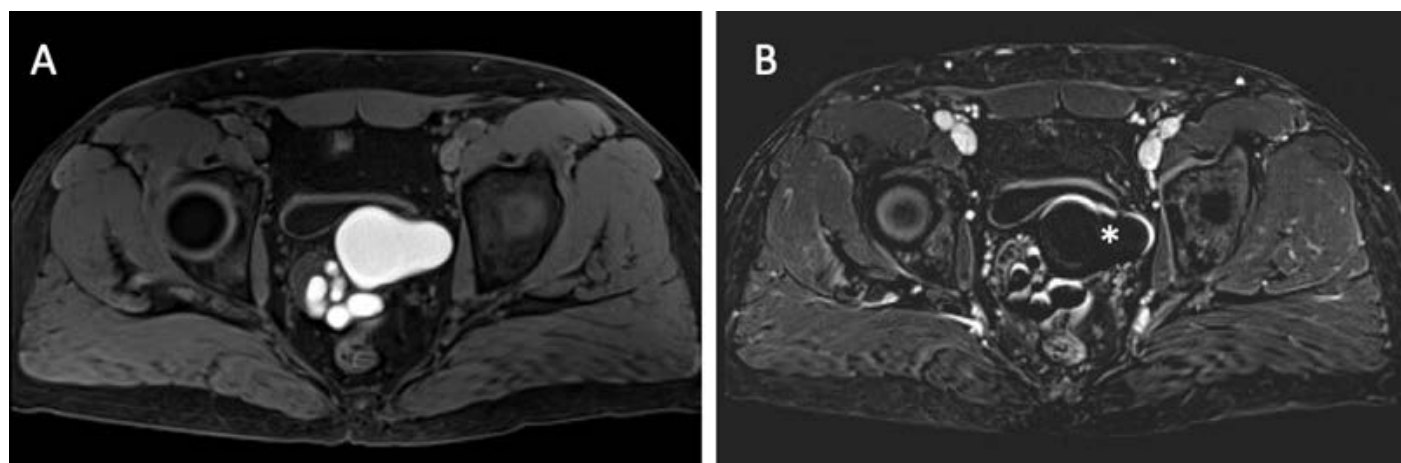


Figure 4: 48-year-old male with pelvic pain and dysuria.

Findings: Axial T1 weighted image with fat saturation reveals the hyperintensity of the cyst and the contralateral vesicle probably due to the presence of proteinaceous content (4A); Axial contrast-enhanced T1 weighted MR subtraction image shows the absence of contrast enhancement (*asterisk*)(4B).

Technique: MRI 1.5 Tesla. 16 mL Gadoteridol (Pro-Hance) MR imaging of the pelvis. A) Axial T1 weighted image with fat saturation. B) Axial contrast-enhanced T1 weighted MR subtraction image.



Figure 5: 48-year-old male with pelvic pain and dysuria.

Findings: Coronal reformatted T1w sequence with MIP reconstruction shows the presence of the superior ectopic atretic ureter (arrow) which drains into the seminal vesicle cyst.

Technique: MRI 1.5 Tesla. 16 mL Gadoteridol (Pro-Hance) MR imaging of the pelvis. Coronal reformatted T1-w sequence with MIP reconstruction.

Etiology	Not fully understood, but it is believed to result from a failure of the Müllerian ducts to regress during fetal development.
Incidence	It is estimated to occur in less than 0.1% of males with only about 200 cases reported in the literature until 2020.
Gender ratio	Only males, since it is considered to be the male equivalent of the Mayer-Rokitansky-Küster-Hauser syndrome.
Age predilection	It is typically diagnosed in early adulthood, with most cases presenting in the third and fourth decades of life.
Risk factors	Not well established due to the limited number of reported cases.
Treatment	Antibiotics in case of urinary tract infection; surgical for large symptomatic cysts.
Prognosis	The long-term prognosis depends on the severity of the symptoms. Most of the time, with timely diagnosis and appropriate treatment, the patients have a good prognosis.

Table 1: Summary table of Zinner's syndrome

Prostatic utricle cyst	Prostatic utricle cysts are embryologic remnants of the Müllerian duct system that communicate with the urethra. They affect predominantly males younger than 20 years. Prostatic utricle cysts always arise from the level of the verumontanum and are always in the midline.
Ejaculatory duct cyst	Ejaculatory duct cysts are rare. They are due to obstruction of the ejaculatory duct that may be congenital or acquired. On imaging, these lesions appear to be cystic structures along the ejaculatory duct just lateral to the midline in the central zone of the prostate.
Prostatic cyst	Retention cysts are acquired cysts that result from obstruction of the glandular ductules, causing dilatation of the acini. They may occur in any glandular zone of the prostate. Retention cysts usually appear as smooth-walled, unilocular simple cysts and rarely cause symptoms.
Diverticula of the ampulla of vas deferens	These cysts are located superior to the prostate gland along the course of the vas deferens. On MRI, vas deferens cysts are easily recognized and distinguished from other adjacent structures.
Ureterocele	Ureteroceles represent abnormal congenital dilatation of the distal-most portion of the ureter. The tortuous course of a hydroureter can mimic a periprostatic cystic lesion. In addition, ectopic insertion of a ureter into the prostatic urethra can resemble a tubular cystic structure when dilated.
Tumor	Both benign and malignant neoplasms may contain cystic components. On MRI, the heterogeneity of signal intensity of the cystic components and the presence of soft-tissue elements within the lesion indicate a neoplastic cause.
Abscess	Abscesses are focal collections of suppurative inflammatory material. Although MRI is usually not performed for this condition, suspicion of an abscess is raised when a cystic lesion with thickened walls, septations, or heterogeneous contents is seen in a patient with appropriate clinical findings.

Table 2: Differential diagnosis table for cystic lesions of the pelvis

ABBREVIATIONS

CT = Computed Tomography; MRI= Magnetic resonance imaging; US = Ultrasound; CAKUT = Congenital anomalies of the kidneys and urinary tracts; DWI = Diffusion weighted imaging; ADC = Apparent diffusion coefficient

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