From page 12

Diagnosis
Infected urachal tract remnant

Overview of the disease
The urachus, or median umbilical ligament, is a midline tubular structure that extends upward from the anterior dome of the urinary bladder toward the umbilicus in the early embryo. The tubular urachus normally involutes during the latter half of fetal life and remains as a fibrous band with no known function. There are four types of congenital urachal anomalies that result if this structure is not obliterated by fibrous proliferation: patent urachus, umbilical-urachal sinus, vesicourachal diverticulum, and urachal cyst. These congenital anomalies are twice as common in men as in women.
The most common anomaly is a patent urachus, accounting for approximately 50 %, and this anomaly is purely congenital. The remaining three entities may be congenital or represent normal closure of the urachus after birth with reopening secondary to acquired pathologic conditions. While the majority of patients with urachal abnormalities (except those with patent urachus) are asymptomatic, symptoms may occur when these are associated with infection or tumor.
Patent urachus is a persistent communication between the bladder lumen and the umbilicus. Urine leakage is usually noted during the neonatal period, and a definitive diagnosis can be made with sinography or cystography. About one-third of patients with a patent urachus have associated posterior urethral valves or urethral atresia.
Umbilical-urachal sinus consists of blind dilatation of the urachus at the umbilicus without communication with the urinary bladder. Periodic discharge may result from the tiny opening into the umbilicus. Infection of the urachal remnant occurs frequently.
Vesicourachal diverticulum is a persistent communication between the urachus and the bladder dome when the vesical end of the urachus fails to close. This lesion may be complicated by urinary tract infection, intraurachal stone formation, and an increased prevalence of carcinoma after puberty. In infants, vesicourachal diverticulum may be accompanied by prune belly syndrome.
Urachal cyst develops if the urachus closes at both the umbilical and vesical ends but remains patent between these endpoints. Urachal cysts primarily occur in the lower one-third of the urachus and vary considerably in size. These become symptomatic when they enlarge or become infected. In fact, the majority of urachal cysts are infected at the time of diagnosis.
Urachal tract remnants that abnormally remain patent are often subject to infection via lymphatic, hematogenous, or vesical routes. A variety of gram-positive and gram-negative micro-organisms have been cultured from infected urachal remnants. These remnants are associated with a 30 % reinfection rate if the cyst wall is not completely removed. Urachal remnants are also associated with benign and malignant neoplasms. Benign urachal tumors including adenomas, fibromas, fibroadenomas, fibromyomas, and hamartomas are extremely rare. Malignant urachal neoplasms are also rare, and the majority of cases are adenocarcinoma (90 %). At histologic analysis, mucin production is found in up to three-fourths of cases. Malignant urachal neoplasms represent less than 0.5 % of all bladder cancers, and 34 % of bladder adenocarcinomas are of urachal origin. These tumors are most commonly seen during the fifth to eighth decades of life, and two-thirds are seen in men. Urachal tumors are typically silent because of their extraperitoneal location, and diagnosis is usually not made until there is local invasion or metastatic disease.

■ RADIOLOGIC OVERVIEW OF THE DIAGNOSIS

Computed tomography and ultrasonography are ideally suited for the detection or urachal remnant diseases. Patent urachus is demonstrated as a tubular connection between the anterosuperior aspect of the bladder and the umbilicus on longitudinal US and occasionally at CT performed during the bladder-filling stage. In the presence of periodic discharge, a thickened tubular structure along the midline below the umbilicus representing an umbilical-urachal sinus can be visualized at US. This can be confirmed at sinography. A vesicourachal diverticulum is demonstrated as a fluid-filled sac protruding from the anterosuperior aspect of the bladder that does not communicate with the umbilicus. In the case of a urachal cyst, CT or US shows a fluid-filled cavity in the midline lower abdominal wall located just beneath the umbilicus or above the bladder. While rare, eggshell calcification of the cyst wall has been reported.
Infected urachal remnants demonstrate complex echogenicity at US and inhomogeneous attenuation with variable contrast enhancement in and around the disease process at CT. Urachal carcinoma may be solid, cystic, or mixed. The US and CT features are similar to that of infected urachal remnants. However, there are a few characteristics that may be used for differentiation. First, calcifications in a midline supravesical mass are considered nearly diagnostic for urachal carcinoma. The calcifications may be punctate, stippled, or curvilinear and peripheral. The lack of adjacent inflammatory change suggests urachal carcinoma. Also, the presence of mural nodularity or hematuria can be helpful as distinguishing features in some cases. Because infected urachal cysts and urachal carcinomas are difficult to differentiate, percutaneous needle biopsy or fluid aspiration is usually needed for diagnosis and therapeutic planning. Urachal carcinomas may be confused with primary tumors of the bladder dome, but the propensity to grow in the perivesical space toward the umbilicus is characteristic of urachal carcinoma. CT may demonstrate metastases initially in the pelvic lymph nodes, followed by systemic metastases to the lung, brain, liver, and bone.

Differential diagnosis

• Mucinous urachal carcinoma
• Nonurachal carcinoma
• Myxoid or mucinous mesenchymal neoplasm
• Bladder metastasis
• Infected vesicourachal diverticulum

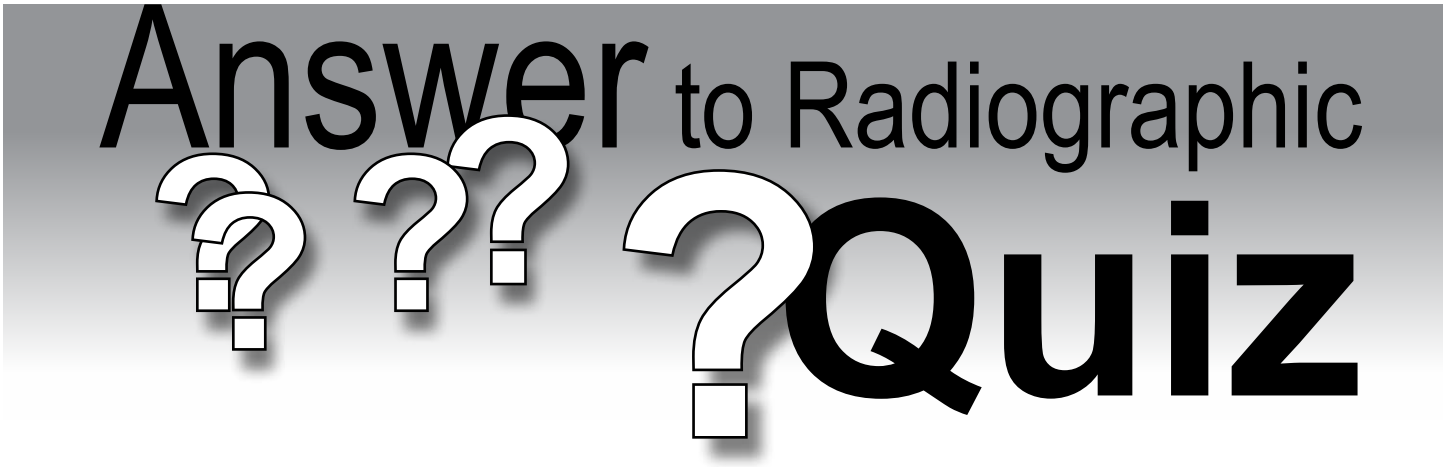
Key Points

• The four types of congenital urachal anomalies are twice as common in men as in women.
• The majority of patients with urachal anomalies (except patent urachus) are asymptomatic.
• Urachal remnants may be associated with infection or neoplasms.
• CT and US findings are similar in infected urachal cyst and urachal carcinoma. Thus, percutaneous needle biopsy or fluid aspiration is often needed for diagnosis.

■ REFERENCES

1. Yu JS, Kim KW, Lee HJ, Lee YJ, Yoon CS, Kim MJ. Urachal Remnant Diseases: Spectrum of CT and US Findings. Radiographics [Internet] 2001; 21:451-461. Available at: https://doi.org/10.1148/radiographics.21.2.g01mr02451

2. Carolina Parada Villavicencio, Sharon Z. Adam, et al. Imaging of the Urachus: Anomalies, complications and mimics. RadioGraphics, 2016. Vol. 36, No. 7. Published Online:Nov 10 2016. Available at: https://doi.org/10.1148/rg.2016160062



Images description

US: Midline supravesical complex mass arising from the dome of the urinary bladder.

CT: Tubular, fluid-filled structure arising from the dome of the urinary bladder without communication with the umbilicus or anterior abdominal wall.

Please answer to the following with ***TRUE*** or ***FALSE*** regarding the salient abnormality and associated findings.

There is an enhancing rim

___*True*
_X_*False*

There is adenopathy

___*True*
_X_*False*

There is bladder invasion

___*True*
_X_*False*

There is free fluid

___*True*
_X_*False*

There is perilesional fat stranding

_X_*True*
___*False*

Where does this mass most likely originate from?

_X_*Bladder*
___*Peritoneum*
___*Small bowel*
___*None of the above*

What is the diagnosis (single best answer, if any)?

___*Ureteritis cystica*
___*Hutch diverticulum*
_X_*Urachal anomaly*
___*Ureteral diverticulum*
___*None of the above*

What is the most likely etiology of this lesion?

_X_ *Infected urachal tract remnants*
___*Umbilical urachal sinus*
___*Vesicourachal diverticulum*
___*Urachal cyst*