Abdominal Radiology (2019) 44:3978–3989
https://doi.org/10.1007/s00261-019-02205-x

SPECIAL SECTION : UROTHELIAL DISEASE

Imaging of urachal anomalies

Suryakala Buddha[1] · **Christine O. Menias**[2] · **Venkata S. Katabathina**[1]

Published online: 3 September 2019

Abstract
Urachal anomalies are classified into four types depending on the level of persistence of the embryonic urachal remnants between the urinary bladder and the umbilicus: patent urachus, umbilical–urachal sinus, urachal cyst, and vesico-urachal diverticulum. Due to the increasing use of cross-sectional imaging, urachal anomalies are frequently detected as incidental findings. Imaging plays a pivotal role in the initial diagnosis, evaluation of complications, treatment follow-up, and long-term surveillance of patients with urachal anomalies. Different urachal anomalies demonstrate characteristic imaging features that aid in a timely diagnosis and guide treatment. A patent urachus is visualized as an elongated tubular structure between the umbilicus and the urinary bladder. While umbilical–urachal sinus appears as focal dilatation at the umbilical end of the urachal remnant, the vesico-urachal diverticulum presents as a focal outpouching of the urinary bladder at anterosuperior aspect. Urachal cysts are identified as midline fluid-filled sacs most frequently located near the dome of the urinary bladder. Untreated urachal anomalies could progress into potential complications, including infection and malignancy. Knowledge regarding imaging features of urachal anomalies helps in timely diagnosis, treatment, follow-up, and early detection of complications.

Keywords Urachus · Imaging · Complications · CT · US · MRI

Introduction

Urachal anomalies develop as the result of incomplete involution and persistence of embryonic urachal remnants after birth [1]. During gestational life, the urachus connects the fetal urinary bladder and the allantois. Usually, at birth, the urachus is obliterated and becomes a vestigial fibromuscular cord-like structure, termed as 'the median umbilical ligament' [1–3]. Postnatally, the median umbilical ligament extends from the dome of the urinary bladder to the umbilicus and lies in the extraperitoneal space of the Retzius, between the parietal peritoneum and the anterior abdominal wall (Fig. 1) [3]. Depending on the location and degree of urachal involution, urachal anomalies can be classified into four types: patent urachus, umbilical–urachal sinus, urachal cyst, and vesico-urachal diverticulum (Fig. 2). Patent urachus (47%) is the most common type, followed by urachal cyst (30%), umbilical–urachal sinus (18%), and least commonly, vesico-urachal diverticulum (3%) [2]. Urachal anomalies may present with clinical symptoms such as umbilical discharge, erythema, mass, and recurrent urinary tract infections due to chronic urinary stasis, which leads to inflammation and infection [4]. Additionally, urachal anomalies can also be identified incidentally in asymptomatic patients with the increased use of cross-sectional imaging [1, 2, 5].

In this article, embryological developmental anatomy of the urachus will be reviewed, and the role of various imaging techniques in the diagnosis and management of patients with urachal anomalies will be discussed. Imaging findings of different urachal anomalies and associated complications such as infection and malignancies will be described.

Embryological development of the urachus

The urachus is formed from the embryological remnants of the allantois and the cloaca (Fig. 3) [6–8]. During 3rd week of the embryonic life, the allantois is formed as an

✉ Venkata S. Katabathina
katabathina@uthscsa.edu

[1] Department of Radiology, University of Texas Health Science Center at San Antonio, Floyd Curl Drive, 7703, San Antonio, TX 78229, USA

[2] Department of Radiology, Mayo Clinic at Scottsdale, Scottsdale, AZ, USA

Fig. 1 Normal CT appearance and anatomical location of the median umbilical ligament (obliterated urachus). Sagittal contrast-enhanced CT images of the abdomen demonstrate the median umbilical ligament (arrows), extending from the dome of the urinary bladder to the umbilicus in the space of Retzius between the anterior abdominal wall and parietal peritoneum

extraembryonic diverticulum from the yolk sac at the tail end of the embryo [2, 3, 6]. During 5th week, with increasing growth and flexure of the embryo, a bend is formed at the junction of hindgut and allantois, which enlarges to become the cloaca [8]. Between 6th and 7th week of embryonic life, the cloaca is divided into anterior primitive urogenital sinus and posterior anorectal canal [8]. The urogenital sinus is further divided into superior pelvic and inferior phallic parts. The superior pelvic portion of urogenital sinus develops into the fetal bladder, and inferior phallic portion develops into urethra [2, 6]. The cephalic extension of the superior part of the urogenital sinus is continuous with the allantois ventrally at the umbilicus; thus, the fetal bladder extends all the way to the umbilicus [2, 3, 6]. During the 4th month of pregnancy as the bladder descends into the pelvis, there is involution of allantois which results in the formation of a tubular structure, the urachus between the apex of the bladder and umbilicus [6]. Approximately between 4th and 5th months of pregnancy and sometimes just before or after birth, the urachus will be obliterated and forms a fibrous cord-like structure that persists as the 'median umbilical ligament' [6].

Role of imaging

A wide array of imaging techniques are helpful in the diagnosis and follow-up of suspected urachal anomalies. They include ultrasonography (US), computed tomography (CT), and magnetic resonance imaging (MRI) [9–11]. In addition, fluoroscopy studies such as voiding cystourethrogram (VCUG), cystogram, and sinogram are also useful in select patients. A sinogram is a special radiographic procedure performed by retrograde injection of water-soluble contrast into the orifice of the urachal tract at the umbilical end. Sinogram helps in the confirmation of a patent urachus and umbilical–urachal sinus [9, 10, 12]. VCUG and cystogram are helpful in the confirmation of vesico-urachal diverticulum and urachal cyst and excludes the presence of vesicoureteral reflux [13]. US is the initial imaging modality as it is widely available, cost-effective, and has a rapid speed of diagnosis. US helps to localize the urachal anomaly, determine its extent and nature (solid vs. cystic) (Fig. 4) [10, 14]. CT and MRI are helpful to evaluate the extent of disease and its potential complications [11]. CT Imaging is rapid and convenient for most patients and useful for disease staging in patients with urachal malignancies [2, 9, 10]. The presence of soft-tissue attenuation lesion in the urachal remnant on unenhanced CT with heterogeneous enhancement after contrast administration suggests the possibility of infection or malignancy (Fig. 5) [3, 9]. MRI has better soft-tissue contrast resolution compared to CT and assists in tissue characterization of associated cystic degeneration changes, and detection of urachal adenocarcinoma [2]. In addition, MRI is helpful in the local staging of urachal tumors and demonstrates their location, extent, and relationship with surrounding structures (Fig. 6) [2].

Patent urachus

Also known as urachal fistula, patent urachus is a persistent tubular connection between the urinary bladder and umbilicus [2]. Posterior urethral valves or urethral atresia can be seen in about one-third of the patients with patent urachus [3]. Most common clinical presentation is seen in the neonatal age with urine leakage from the umbilicus leading to umbilical edema and delayed cord stump healing [2, 8]. If communication is extremely narrow, patients may be asymptomatic. Children and adults may also present with urine leakage or purulent discharge from the umbilicus, with painful, palpable suprapubic mass if the tract gets infected [7].

On US, a patent urachus appears as a tubular fluid-filled structure between the anterosuperior aspect of the bladder and umbilicus with a hypoechoic wall and anechoic lumen (Fig. 7) [2, 3]. A confident diagnosis of patent urachus can

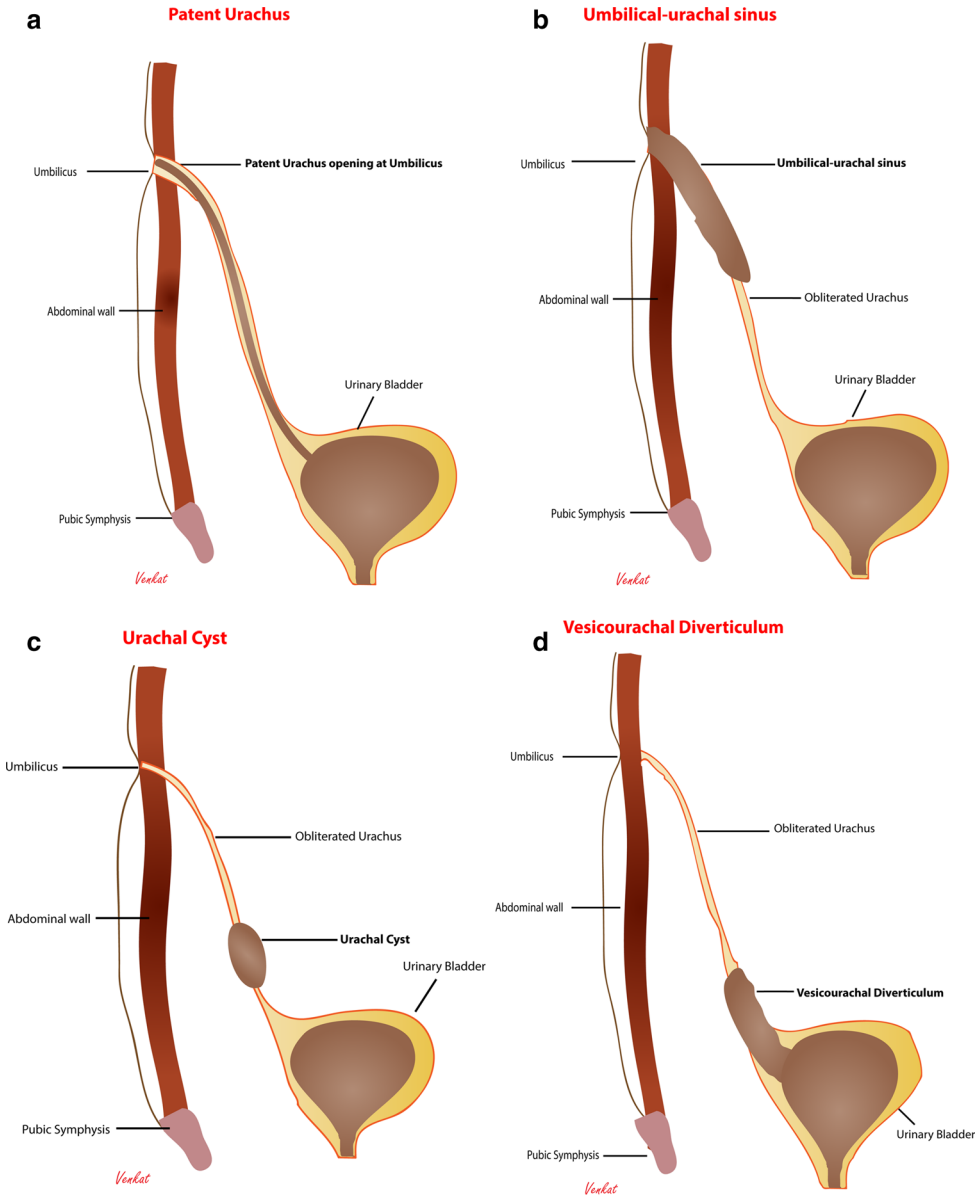

Fig. 2 Illustrations demonstrate various types of urachal anomalies. **a** Patent urachus, a persistent tubular connection between the urinary bladder and umbilicus. **b** Umbilical–urachal sinus, a fusiform dilatation of the urachus at the umbilical end. **c** Urachal cyst, a cystic lesion along the course of the median umbilical ligament. **d** Vesico-urachal diverticulum, an outpouching arising from the anterosuperior aspect of the urinary bladder

be made with a sinogram or VCUG; in these fluoroscopic studies, the presence of a contrast-filled tubular structure between the urinary bladder and the umbilicus confirms the diagnosis of patent urachus (Fig. 7) [2, 3, 5]. Additionally, VCUG can help exclude posterior urethral valves and vesicoureteral reflux [2]. Contrast-enhanced CT images demonstrate a tubular structure containing air spanning from the dome of the urinary bladder to the umbilicus with variable enhancement [3]. On MRI, an uncomplicated patent urachus appears as a T1-hypointense, T2-hyperintense tubular tract without significant enhancement [2].

Umbilical–urachal sinus

The umbilical–urachal sinus develops due to failure of involution of the urachus resulting in the fusiform dilatation of the urachus at the umbilical end [2]. Cystic dilation of the urachus that empties periodically into the umbilicus or the urinary bladder is known as ‘alternating sinus’ [5]. The umbilical–urachal sinus may be associated with urachal cyst and probably could be due to sequelae of an infected urachal cyst that drained at the umbilicus [2, 5]. The umbilical–urachal sinus has a potential space that allows the accumulation of cellular debris and superimposed infection, and rarely stone formation [2]. Clinically, patients present with periumbilical inflammation with pain or mass with or without purulent discharge [5, 13].

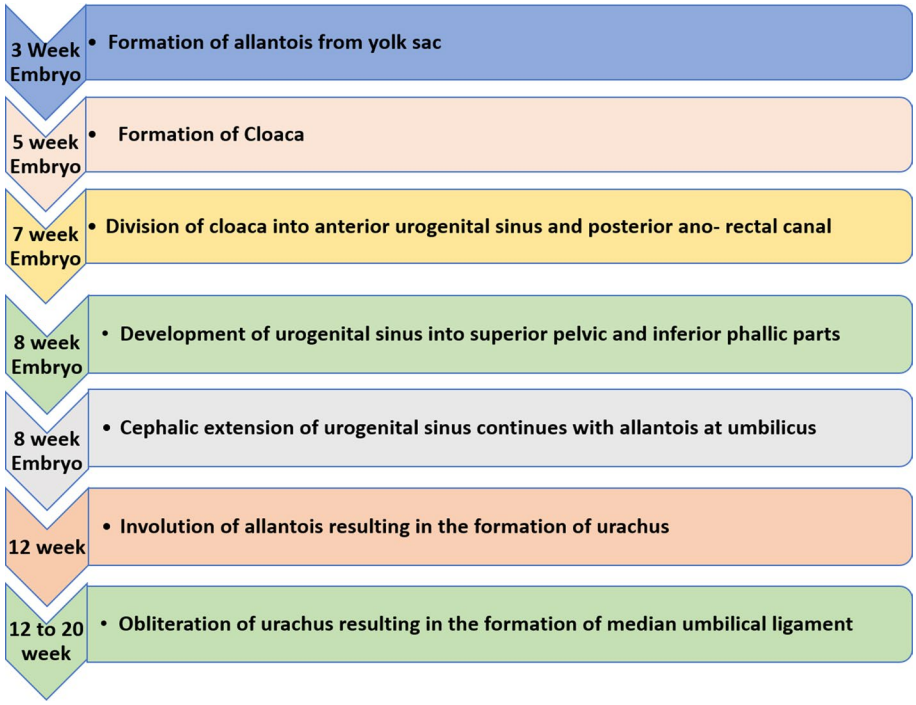

Fig. 3 A flowchart that demonstrates embryological development of the urachus

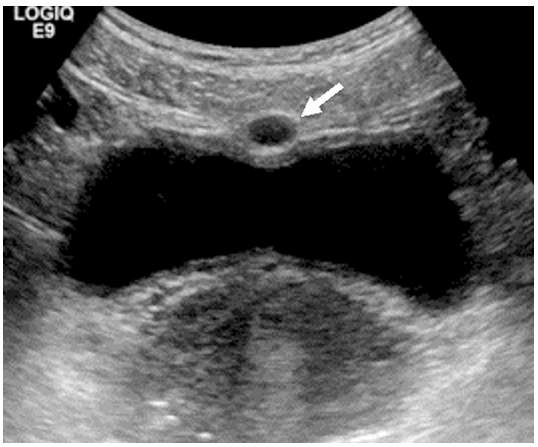

Fig. 4 A 17-year-old woman with an urachal cyst. Transverse gray-scale US image demonstrates a well-defined, ovoid anechoic cystic lesion abutting the dome of the urinary bladder without any definite communication (arrow), suggestive of an urachal cyst

US demonstrates a thickened hypoechoic tubular structure spanning from the umbilicus just below the anterior abdominal wall without a definite communication with the urinary bladder [2]. Sinogram is the confirmatory study for the diagnosis of the umbilical–urachal sinus; and demonstrates a blind-ending tract originating from the umbilicus without communication to the urinary bladder [1, 8]. CT and MRI help in excluding superimposed infection and malignancy of the umbilical–urachal sinus [2, 3].

Fig. 5 An infected vesico-urachal diverticulum in a 35-year-old man with recurrent urinary tract infections. Axial contrast-enhanced CT image shows a vesico-urachal diverticulum (arrow) in the anterosuperior aspect of the urinary bladder with adjacent fat stranding, suggestive of an infected vesico-urachal diverticulum

Urachal cyst

An urachal cyst develops if the umbilical and vesical ends of the urachus obliterate, and the intervening lumen remains patent forming the cystic lesion [3, 6]. Urachal cyst commonly occurs in the lower one-third of the urachus, in the proximity to the urinary bladder [2, 3]. Usually, the urachal cysts are small in size and are

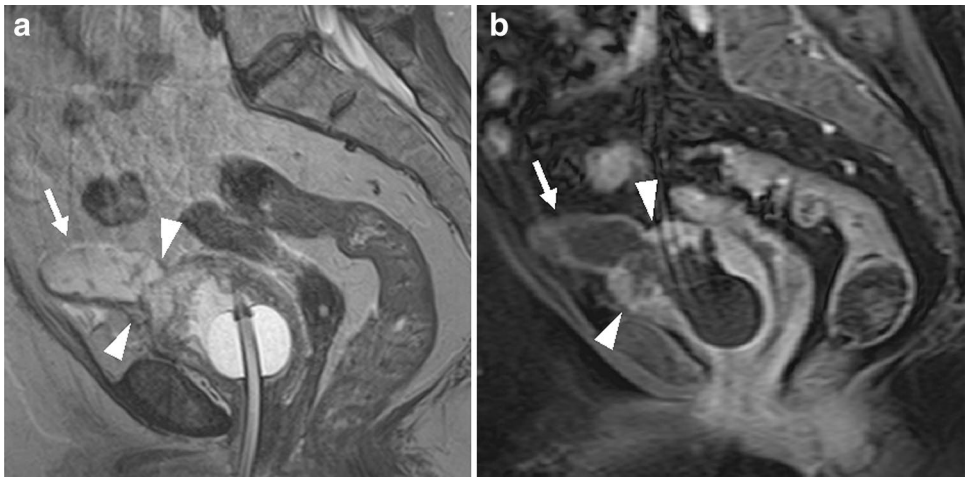

Fig. 6 Urachal adenocarcinoma in a 51-year-old woman with vesico-urachal diverticulum. Sagittal T2-weighted (**a**) and contrast-enhanced T1-weighted (**b**) MR images of the pelvis demonstrate a vesico-urachal diverticulum (arrow) at the anterosuperior aspect of the urinary bladder with irregular wall thickening and heterogeneous enhancement, concerning for malignancy (arrowheads). This was proven to be a vesico-urachal diverticulum with adenocarcinoma on pathology

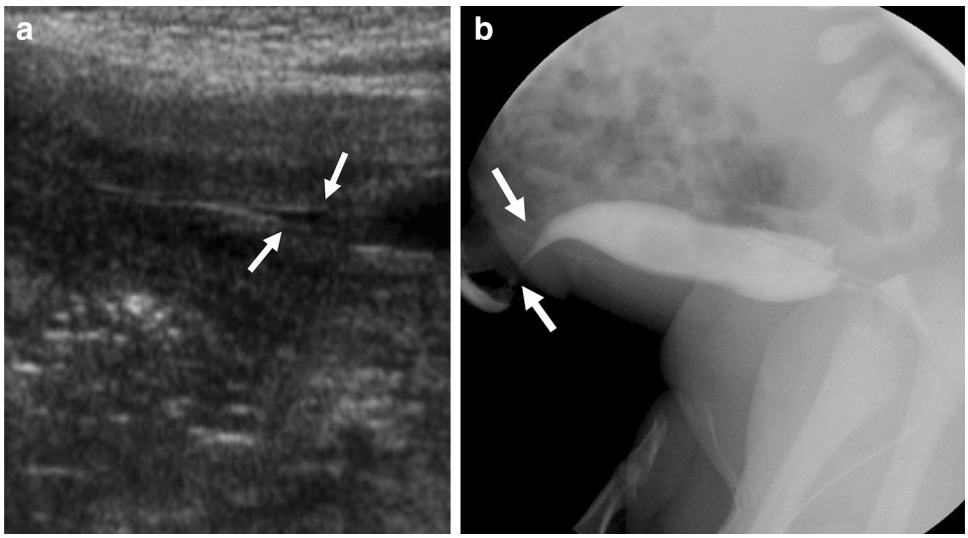

Fig. 7 An infant with a patent urachus. Longitudinal gray-scale US image **a** shows an anechoic, tubular fluid-filled structure (arrows) between the umbilicus and urinary bladder (not shown). A lateral view spot radiograph obtained during voiding cystourethrography **b** shows a contrast-filled fistulous tract (arrows) between the umbilicus and the urinary bladder

asymptomatic and detected incidentally at imaging performed for the other indications [3]. However, patients may also present with abdominal pain due to hemorrhage into the cyst or spontaneous rupture of an uninfected cyst or intraabdominal rupture of the infected cyst leading to peritonitis [2, 3].

On US, urachal cyst appears as a thin-walled, midline cystic lesion with posterior acoustic enhancement along the course between the umbilicus and the urinary bladder, commonly abutting the urinary bladder dome (Fig. 4) [2, 3, 8]. VCUG excludes the communication of the cyst with urinary bladder [3, 8]. At CT, urachal cyst presents as an extraperitoneal, non-communicating, midline cystic lesion between the umbilicus and anterosuperior aspect of the bladder [2, 3]. MRI is indicated for better evaluation of the cyst and its relation with the urinary bladder. On MRI, urachal cyst appears as a thin-walled homogenous, T1-hypointense, T2-hyperintense, non-enhancing cystic lesion (Fig. 8) [2, 3, 5]. The common complications of the urachal cyst are infection, the

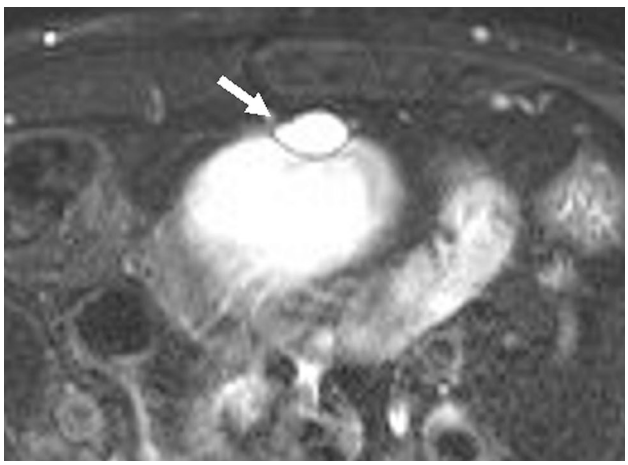

Fig. 8 A 32-year-old man with an urachal cyst. Axial T1-weighted MR image shows a well-defined T2 hyperintense cyst (arrow), abutting the anterior and superior surface of the urinary bladder consistent with an urachal cyst

formation of wall calcifications, stones, and rupture into the peritoneal cavity leading to peritonitis [1, 3, 15].

Vesico-urachal diverticulum

Vesico-urachal diverticulum develops when the supra-vesical portion of the urachus fails to involute completely resulting in the formation of an outpouching of variable length from the anterosuperior aspect of the urinary bladder, which does not communicate with the umbilicus [2, 3]. Vesico-urachal diverticulum is a rare urachal anomaly that represents 3% of all urachal anomalies and is usually asymptomatic in adults [5]. In infants, it is commonly accompanied by prune-belly syndrome [3, 12].

On US, vesico-urachal diverticulum appears as an extraluminal, hypoechoic outpouching arising in the anterosuperior aspect of the bladder, not communicating with the umbilicus (Fig. 9) [1–3, 8]. During VCUG examination, contrast reflux is seen in the vesico-urachal diverticulum, and it appears as a contrast-filled structure extending from the anterosuperior aspect of the bladder even after voiding (Fig. 9) [3, 16]. CT/MRI studies demonstrate a heterogeneously enhancing, blind-ending soft-tissue tract and/or an irregular mass in the space of Retzius at the anterosuperior aspect of the urinary bladder (Figs. 5, 6, 10) [2, 3]. Complications of vesico-urachal diverticulum include recurrent urinary tract infections, chronic bladder outlet obstruction, intra-diverticular calculi from urinary stasis, and increased risk of urachal neoplasms in adults (Figs. 5, 6, 10) [7, 12].

Fig. 10 A 64-year-old man with a vesico-urachal diverticulum containing a calculus. Contrast-enhanced sagittal CT image of the abdomen shows a vesico-urachal diverticulum (arrow) containing a calculus (arrowhead)

Management of urachal anomalies

The treatment approach for urachal anomalies remains controversial and depends upon the patient's age and severity

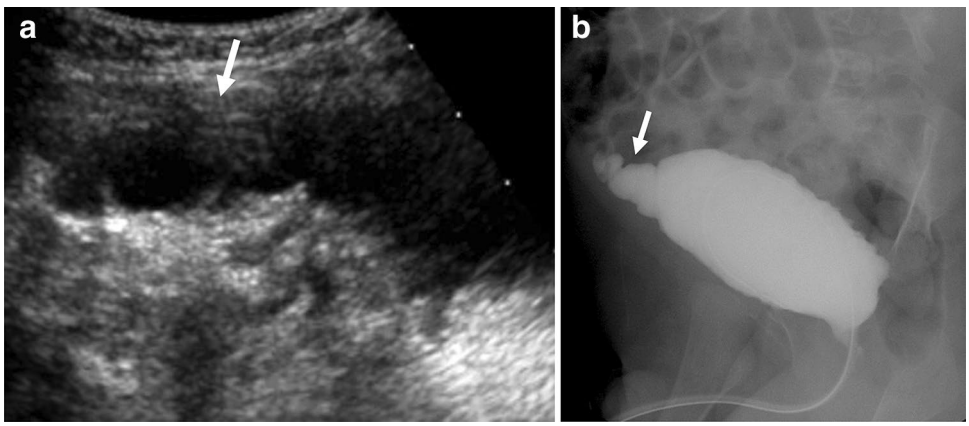

Fig. 9 A 7-year-old boy with vesico-urachal diverticulum. **a** Gray-scale US image of the pelvis demonstrates an anechoic, outpouching (arrow) from the anterosuperior aspect of the urinary bladder (arrowhead), suggestive of a diverticulum. **b** A lateral view spot radiograph obtained during voiding cystourethrography shows a contrast-filled outpouching (arrow) arising from the anterosuperior aspect of the urinary bladder consistent with a vesico-urachal diverticulum

of the symptoms [1–3]. Although surgical management is required in the majority of the symptomatic patients, asymptomatic individuals, patients with non-specific atretic urachal remnants, and select symptomatic patients can safely be managed non-operatively [10, 17]. With increasing case reports of spontaneous resolution of urachal anomalies after birth, symptomatic patients are first managed medically and only in select patients without spontaneous resolution 1 year after birth are managed surgically [18]. Follow-up ultrasound is recommended to evaluate for any evidence of urachal remnants in these patients [19].

Prophylactic surgical excision of asymptomatic urachal anomalies in adults is recommended to prevent potential complications such as infection and malignancy [10]. However, this approach is too aggressive in children as the possibility of malignancy is very rare [12]. Complete excision of the urachal remnants is recommended for all urachal anomalies, either by open or laparoscopic surgical approach [10, 20, 21]. Urachal anomalies that involve the bladder (patent urachus and vesico-urachal diverticulum) requires excision of remnant tissue along with placement of urinary bladder cuff to prevent long-term complications [14, 21]. This radical excision includes excision of the medial umbilical ligaments along with urachal remnants and adjacent peritoneum [14, 20, 21].

Complications

The development of complications in patients with urachal anomalies depends on the patient's age and the type of urachal anomaly [2, 3]. While infection of the urachal remnants is common in infants and children, infection and malignancy are more common in adults [2, 3].

Infection

Infection of urachal remnant tissue is the most common complication of urachal anomalies [3, 8]. The route of infection could be hematogenous, lymphatic, and direct extension from the urinary bladder. Predisposing factors for infection, especially in infants and children, include vesicoureteric reflux, umbilical hernia, hypospadias, cryptorchidism, anal atresia, omphalocele, and ureteropelvic junction obstruction [3]. Infection can be caused by gram-positive and gram-negative organisms, with *Staphylococcus aureus* being the most common organism [4, 9, 22]. Clinically, patients present with fever, abdominal pain, and tenderness. Drainage of infected fluid depends on the type of urachal anomaly [2]. Intraperitoneal rupture of the urachal cyst may lead to localized or generalized peritonitis, sepsis, abscess, and fistula formation [3, 15, 23].

US, CT, and MRI can be used for the evaluation of infected urachal remnants [13]. While US is the first screening imaging tool in suspected cases of infection, CT and MRI are used as definite confirmatory imaging modalities and to evaluate the anatomic relation of the infected urachal anomaly with adjacent structures [2, 15, 22, 24]. On US, echogenic contents with irregular wall thickening within the urachal anomaly and a complex, heterogeneous echogenic mass between the dome of the urinary bladder and umbilicus raise the suspicion of infection [2, 3]. CT and MRI demonstrate heterogeneous attenuation/signal intensity with variable contrast enhancement (Figs. 5, 11)

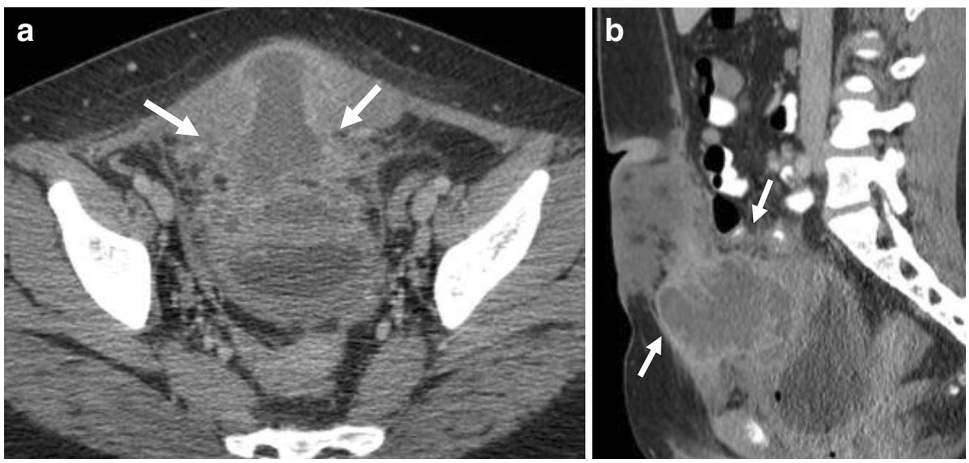

Fig. 11 Infected urachal cyst in a 22-year-old woman. Contrast-enhanced axial (**a**) and sagittal (**b**) images of the abdomen show an ill-defined fluid collection in the anterosuperior aspect of the urinary bladder with peripheral rim enhancement (arrows) and soft-tissue edema in the abdominal wall, extending up to the level of the umbilicus. This patient underwent percutaneous drainage and subsequent surgical excision after the resolution of inflammation. This lesion was proven to be an infected urachal cyst on pathology

[2]. MRI helps in better tissue characterization, evaluation of the extent of infection along with the anatomical relationship of urachus with the urinary bladder, and anterior abdominal wall [5, 13]. There is a 30% risk of recurrent infection and malignant degeneration secondary to incomplete removal of cyst wall or urachal remnants [3, 5]. The presence of heterogeneous echogenicity within the cystic lesion on US examination suggests the possibility of infection in the urachal cyst [3]. At CT/MRI, irregular wall thickening, heterogeneous enhancement, and adjacent fat stranding indicate infected urachal cyst (Fig. 11) [2, 3, 5, 8].

Urachal neoplasms

Both benign and malignant tumors can develop from urachal remnants. Benign urachal tumors are extremely rare and include adenomas, fibromas, fibroadenomas, hamartomas, lipomas, and inflammatory pseudotumors (Fig. 12) [2, 25].

Malignant urachal tumors are also very rare and constitute 1% of all urinary bladder malignancies [2, 26]. Approximately 80% of urachal carcinomas are adenocarcinomas, both mucin-producing and non-mucinous tumors; urothelial, squamous, and sarcomatoid malignancies represent the remaining 20% [2, 25, 27, 28]. Most urachal carcinomas originate at the juxtavesical location and grow cranially toward the umbilicus and inferiorly toward the urinary bladder. Adenocarcinomas arising in urachal remnants are likely due to chronic urinary stasis and result in columnar metaplasia of the transitional epithelium [2, 29]. Majority of the urachal carcinomas are clinically silent due to an extraperitoneal location in the space of Retzius. Patients usually present in later stages of the disease with local invasion to surrounding structures and distant metastases [2]. Hematuria and a presenting palpable suprapubic mass are the most common symptoms; other presentations include abdominal pain, dysuria, bloody discharge from the umbilicus, and obstructive urinary symptoms [27].

Diagnostic evaluation of urachal carcinomas includes clinical history, physical exam, cystoscopy, and urine analysis with urinary cytology [3]. Urachal carcinomas are typically large masses and manifest with prominent extra-vesical component compared to the non-urachal tumors of the urinary bladder [3]. Cystoscopy helps to directly visualize the tumor in the bladder and determine if the tumor has intact mucosa or ulcerated lesion [30]. The dedicated US is the initial imaging modality that confirms the location and extent of the tumor, presence of pelvic lymph nodes, and distant metastases [2, 26]. On US, urachal carcinoma appears as a soft-tissue mass at the urinary bladder dome adjacent to the lower abdominal wall with heterogeneous echogenicity and increased vascularity (Fig. 13) [5, 25, 27, 31].

At CT/MRI, urachal carcinomas may present with a variety of imaging appearances; tumor can present as a large, heterogeneous necrotic, midline mass adjacent to the dome of the urinary bladder, which can extend into the space of Retzius, and demonstrate focal irregular wall thickening, or a small, enhancing nodule near the bladder dome (Figs. 13, 14, 15) [2, 12, 25, 27]. The presence of calcifications (due to mucinous differentiation) is pathognomonic of urachal adenocarcinomas and can be seen in up to 70% of cases at the periphery of the mass (Fig. 14) [2, 3]. On MRI, tumors show areas of high signal intensity on T2-weighted images, which is likely due to mucin content of the tumor or necrosis [27, 32]. The solid component of the tumor demonstrates heterogeneous enhancement after intravenous contrast

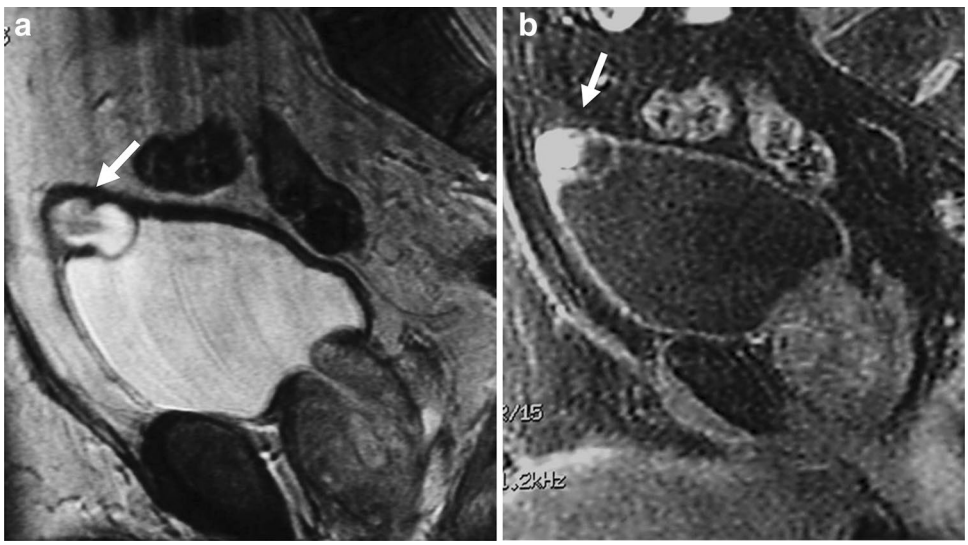

Fig. 12 A 72 year-old-man with tubulovillous adenoma in a vesico-urachal diverticulum. Sagittal T2-weighted (**a**) and contrast-enhanced sagittal T1-weighted (**b**) MR images demonstrate an ill-defined T2 hypointense mass within the vesico-urachal diverticulum with homogeneous enhancement after contrast administration (arrows). This mass was proven to be a tubulovillous adenoma in the vesico-urachal diverticulum on pathology

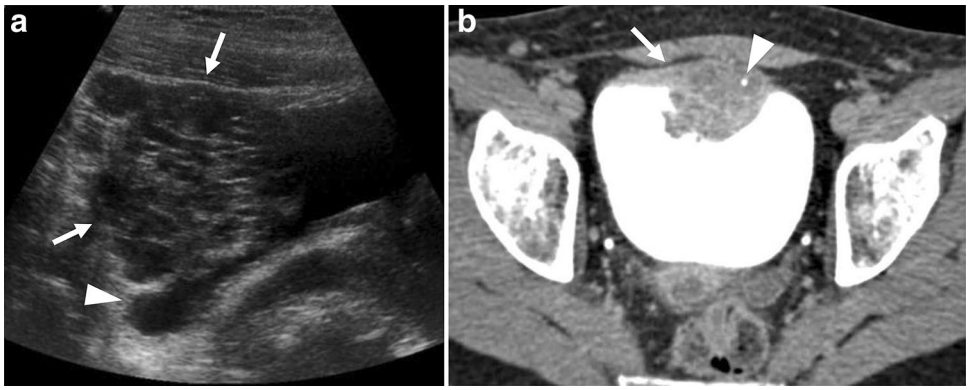

Fig. 13 A 45-year-old man with an urachal adenocarcinoma. **a** Transverse grayscale US image of the pelvis shows an ill-defined heterogeneously hypoechoic mass (arrows) causing significant compression on the urinary bladder (arrowhead). **b** Contrast-enhanced CT image obtained during the excretory phase of CT urogram study demonstrates a heterogeneously enhancing soft-tissue attenuation mass (arrow) arising from the anterior wall of the urinary bladder with a punctate focus of calcification (arrowhead). This mass was proven to be an urachal adenocarcinoma on pathology

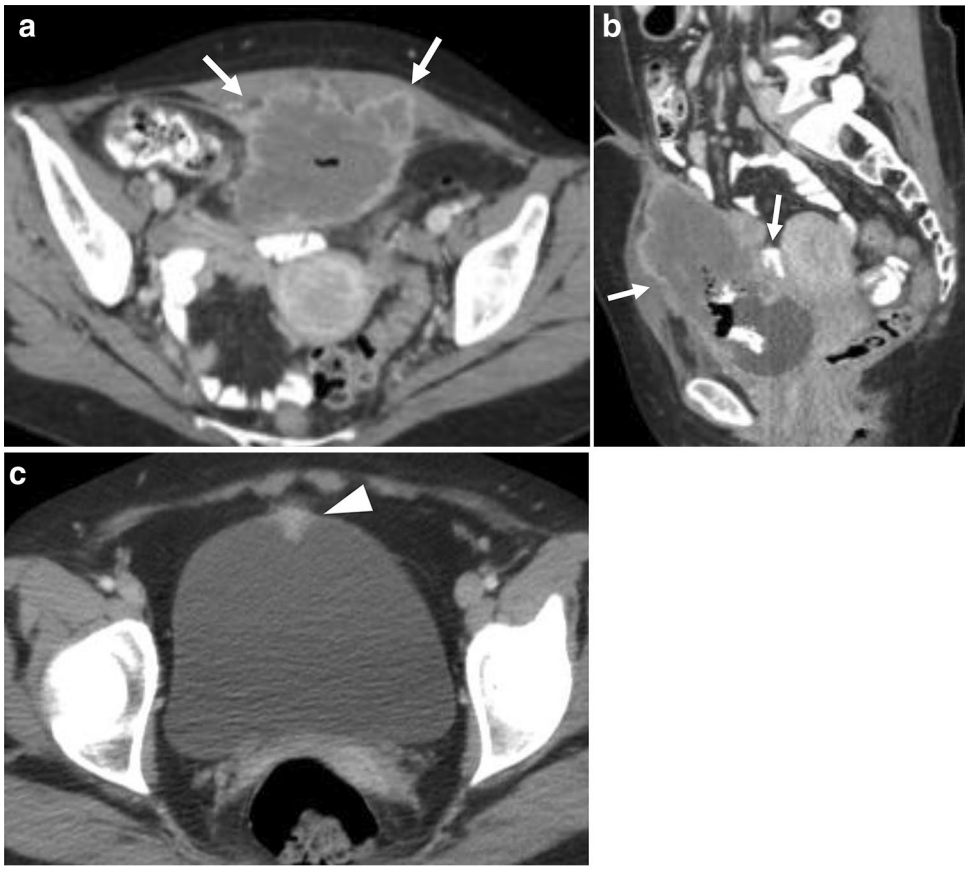

Fig. 14 A 45-year-old woman with an urachal adenocarcinoma. **a**, **b** Contrast-enhanced axial (**a**) and sagittal (**b**) images of the pelvis demonstrate an ill-defined, peripherally enhancing hypoattenuating mass (arrows) with foci of air and peripheral calcifications arising from the dome of the urinary bladder. **c** Contrast-enhanced axial CT image of the pelvis performed 5 years ago demonstrates focal wall thickening and enhancement (arrowhead) in the dome of the urinary bladder that was missed initially. This mass was proven to be an urachal adenocarcinoma on surgical excision

administration [27]. Urachal carcinomas are better identified on sagittal CT/MRI images with adequately distended urinary bladder.

Multiple clinical and imaging staging systems for urachal carcinomas have been proposed, but none of them have been validated so far [2, 28]. Sheldon pathologic staging is the first proposed staging system for urachal cancers; however, this was replaced by Mayo clinic staging proposed by Ashely et al. and Ontario staging system proposed by Pinthus et al. [33–35]. The TNM staging of urachal carcinoma assesses the extent of the disease such as tumor localized to bladder wall or metastasis to lymph nodes or distant organs [8, 28].

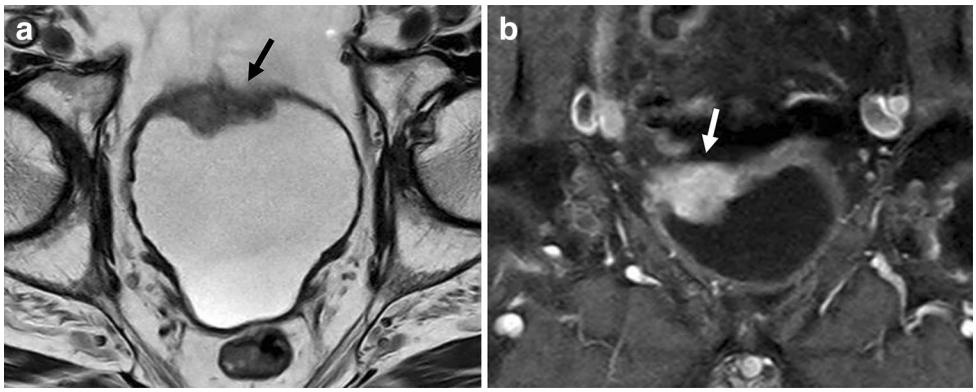

Fig. 15 An 80-year-old man with urachal adenocarcinoma. Axial T2-weighted (**a**) and contrast-enhanced coronal T1-weighted (**b**) MR images show focal wall thickening and enhancement of the urinary bladder dome (arrows), which was proven to be an urachal adenocarcinoma on surgical excision

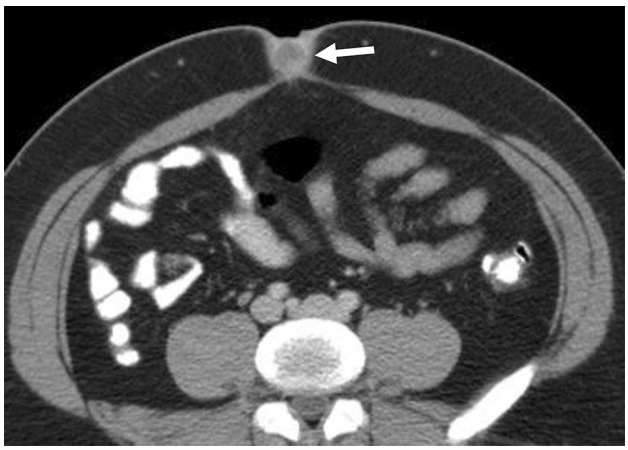

Fig. 16 Umbilical abscess mimicking an umbilical–urachal sinus. Contrast-enhanced axial CT image of the abdomen shows a hypoattenuating focal lesion with rim enhancement in the umbilicus (arrow). This was proven to be an umbilical abscess without any urachal remnants on surgical excision. However, this mimics umbilical–urachal sinus on imaging

Imaging studies play a pivotal role in the initial diagnosis, staging, treatment follow-up, and long-term surveillance of urachal carcinomas [2, 5]. Tumors commonly metastasize initially to pelvic lymph nodes, followed by systemic metastasis to the brain, liver, lungs bowel, and bones [5, 26].

Radical cystectomy, en-bloc resection of the urachal mass, urachal remnants, umbilicus, and pelvic lymph nodes is the treatment of choice for locally advanced urachal carcinomas [36, 37]. In case of advanced urachal cancers, taxanes with a regimen to treat the gastrointestinal malignancy is used due to the enteric type of histology of adenocarcinoma. Hence, the combination of gemcitabine, cisplatin, and 5-fluorouracil has shown to slow down the disease in metastatic urachal carcinoma. A recent study has shown that cytoreductive surgery, and hyperthermic intraperitoneal chemotherapy (HIPEC) prolongs survival time in patients with peritoneal metastases and urachal carcinoma [38]. The prognosis of urachal carcinomas is poor due to their late presentation and advanced stage with local invasion. However, the prognosis is significantly better compared to urinary bladder urothelial carcinoma at similar stages likely due to a relatively younger age of presentation of urachal carcinoma. The 5-year survival rate for urachal carcinoma varies and is reported as 49% after treatment [2, 36]. Approximately 90% of patients presenting with metastases die within 1 year of presentation [34, 39].

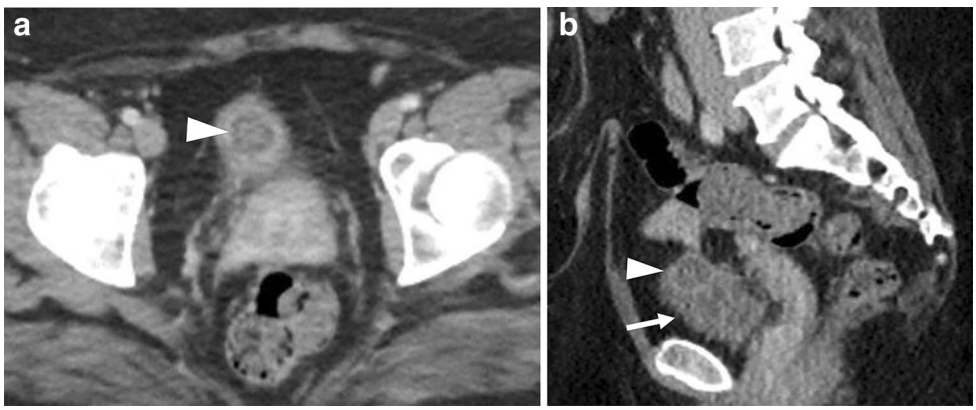

Fig. 17 A 63-year-old woman with a dropped gall stones on the urinary bladder dome mimicking an urachal adenocarcinoma. **a**, **b** Contrast-enhanced axial (**a**) and sagittal (**b**) CT images of the pelvis demonstrate a collapsed urinary bladder (arrow) with a dropped gall stone abutting the urinary bladder dome (arrowheads) mimicking an urachal adenocarcinoma

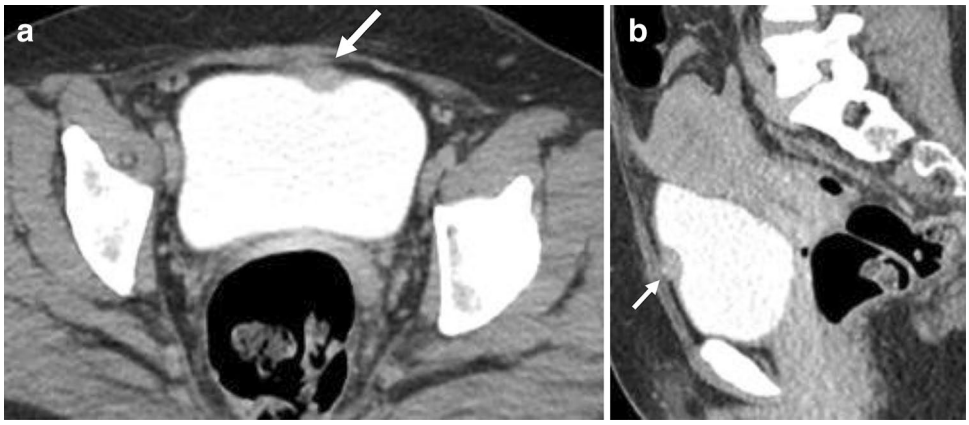

Fig. 18 A 38-year-old woman with a focal endometriotic deposit on the anterior wall of the urinary bladder mimicking an urachal adenocarcinoma. **a**, **b** Contrast-enhanced axial (**a**) and sagittal (**b**) CT images of the pelvis obtained during the excretory phase of CT urogram study demonstrate a well-defined, enhancing mass in the anterior wall of the urinary bladder (arrows). This was proven to be a focus of endometriosis on partial cystectomy, however, mimicking an urachal adenocarcinoma on imaging

Mimics of urachal anomalies

Multiple disease entities involving the periumbilical region, space of the Retzius, and those abutting the urinary bladder dome may simulate urachal anomalies. Umbilical–urachal sinus may mimic omphalitis with umbilical abscess and umbilical granulomas (Fig. 16). Uncomplicated urachal cysts may present similar to urinary bladder diverticulum, mesenteric cyst, Meckel's diverticulum, and umbilical hernia [2]. A vesico-urachal diverticulum can be confused with urinary bladder diverticulum and post-surgical fluid collections. Urachal carcinomas mimic dropped gall stones, endometriosis, and infected urachal remnants (Figs. 17, 18) [2]. Urachal carcinomas also mimic malignant tumors such as urothelial bladder cancer, and omental metastases that abut the bladder dome [2, 3]. Imaging studies, especially contrast-enhanced CT and MRI, help in achieving an appropriate diagnosis.

Conclusion

Depending on the level of persistence of the embryonic urachal remnants between the urinary bladder and the umbilicus, urachal anomalies can be classified into four types which include patent urachus, umbilical–urachal sinus, urachal cyst, and vesico-urachal diverticulum. Early identification of urachal anomalies is crucial to optimize appropriate medical management and if required, surgical resection of the remnants to prevent potential complications, which include infection and malignancy. Select urachal anomalies demonstrate characteristic imaging findings and imaging studies play an essential role in the diagnosis, follow-up, and surveillance of patients with urachal anomalies. Knowledge and understanding of the embryological anatomy of the urachus, along with typical locations and imaging features of urachal anomalies is essential for accurate diagnosis and proper management.

References

1. Fahmy M (2018) Urachal Anomalies. In: Fahmy M (ed) Umbilicus and Umbilical Cord, vol 10. Springer International Publishing, Cham, pp 229–252. https://doi.org/10.1007/978-3-319-62383-2_35
2. Parada Villavicencio C, Adam SZ, Nikolaidis P, Yaghmai V, Miller FH (2016) Imaging of the Urachus: Anomalies, Complications, and Mimics. Radiographics 36 (7):2049-2063. https://doi.org/10.1148/rg.2016160062
3. Yu JS, Kim KW, Lee HJ, Lee YJ, Yoon CS, Kim MJ (2001) Urachal remnant diseases: spectrum of CT and US findings. Radiographics 21 (2):451-461. https://doi.org/10.1148/radiographics.21.2.g01mr02451
4. Ashley RA, Inman BA, Routh JC, Rohlinger AL, Husmann DA, Kramer SA (2007) Urachal anomalies: a longitudinal study of urachal remnants in children and adults. The Journal of urology 178 (4 Pt 2):1615-1618. https://doi.org/10.1016/j.juro.2007.03.194
5. Khati NJ, Enquist EG, Javitt MC (1998) Imaging of the umbilicus and periumbilical region. Radiographics: a review publication of the Radiological Society of North America, Inc 18 (2):413-431. https://doi.org/10.1148/radiographics.18.2.9536487
6. Elumalai G AB (2017) Congenital Anomalies of Urachus"-Embryological basis and its clinical significance.:45676–46679
7. Grosfeld JL (2006) Pediatric surgery. 6th ed./[edited by] Jay L. Grosfeld… [et al.] edn. Mosby Elsevier, Philadelphia, Pa.; Edinburgh
8. V. S. Teran Pareja AMA,; N. Guerrero Salcedo,; E. Prado Miranda,; E. Rico Aragón,; E. Martínez Carapeto; Madrid/ES. (2015) Clinical and Radiological Features of congenital and acquired urachal remnant diseases. https://doi.org/10.1594/ecr2015/c-2538

9. Cilento BG, Jr., Bauer SB, Retik AB, Peters CA, Atala A (1998) Urachal anomalies: defining the best diagnostic modality. Urology 52 (1):120-122
10. Naiditch JA, Radhakrishnan J, Chin AC (2013) Current diagnosis and management of urachal remnants. Journal of pediatric surgery 48 (10):2148-2152. https://doi.org/10.1016/j.jpedsurg.2013.02.069
11. Yiee JH, Garcia N, Baker LA, Barber R, Snodgrass WT, Wilcox DT (2007) A diagnostic algorithm for urachal anomalies. Journal of pediatric urology 3 (6):500-504. https://doi.org/10.1016/j.jpurol.2007.07.010
12. Gleason JM, Bowlin PR, Bagli DJ, Lorenzo AJ, Hassouna T, Koyle MA, Farhat WA (2015) A comprehensive review of pediatric urachal anomalies and predictive analysis for adult urachal adenocarcinoma. The Journal of urology 193 (2):632-636. https://doi.org/10.1016/j.juro.2014.09.004
13. Nimmonrat A, Na-ChiangMai W, Muttarak M (2008) Urachal abnormalities: clinical and imaging features. Singapore medical journal 49 (11):930-935
14. Little DC, Shah SR, St Peter SD, Calkins CM, Murphy JP, Gatti JM, Gittes GK, Sharp RJ, Andrews WS, Holcomb GW, 3rd, Ostlie DJ, Snyder CL (2005) Urachal anomalies in children: the vanishing relevance of the preoperative voiding cystourethrogram. Journal of pediatric surgery 40 (12):1874-1876. https://doi.org/10.1016/j.jpedsurg.2005.08.029
15. Choi YJ, Kim JM, Ahn SY, Oh J-T, Han SW, Lee JS (2006) Urachal Anomalies in Children: A Single Center Experience. Yonsei medical journal 47 (6):782. https://doi.org/10.3349/ymj.2006.47.6.782
16. Ozbülbül NI, Dağli M, Akdoğan G, Olçer T (2010) CT urography of a vesicourachal diverticulum containing calculi. Diagnostic and interventional radiology (Ankara, Turkey) 16 (1):56–58. https://doi.org/10.4261/1305-3825.dir.1798-08.1
17. Lipskar AM, Glick RD, Rosen NG, Layliev J, Hong AR, Dolgin SE, Soffer SZ (2010) Nonoperative management of symptomatic urachal anomalies. Journal of pediatric surgery 45 (5):1016–1019. https://doi.org/10.1016/j.jpedsurg.2010.02.031
18. Galati V, Donovan B, Ramji F, Campbell J, Kropp BP, Frimberger D (2008) Management of urachal remnants in early childhood. The Journal of urology 180 (4 Suppl):1824-1826; discussion 1827. https://doi.org/10.1016/j.juro.2008.03.105
19. Nogueras-Ocaña M, Rodríguez-Belmonte R, Uberos-Fernández J, Jiménez-Pacheco A, Merino-Salas S, Zuluaga-Gómez A (2014) Urachal anomalies in children: Surgical or conservative treatment? Journal of pediatric urology 10 (3):522–526. https://doi.org/10.1016/j.jpurol.2013.11.010
20. Bertozzi M, Riccioni S, Appignani A (2014) Laparoscopic Treatment of Symptomatic Urachal Remnants in Children. Journal of Endourology 28 (9):1091–1096. https://doi.org/10.1089/end.2014.0203
21. Araki M, Saika T, Araki D, Kobayashi Y, Uehara S, Watanabe T, Yamada K, Nasu Y, Kumon H (2012) Laparoscopic management of complicated urachal remnants in adults. World journal of urology 30 (5):647-650. https://doi.org/10.1007/s00345-012-0829-x
22. Yoo KH, Lee S-J, Chang S-G (2006) Treatment of Infected Urachal Cysts. Yonsei medical journal 47 (3):423. https://doi.org/10.3349/ymj.2006.47.3.423
23. Spataro RF, Davis RS, McLachlan MS, Linke CA, Barbaric ZL (1983) Urachal abnormalities in the adult. Radiology 149 (3):659-663. https://doi.org/10.1148/radiology.149.3.6647841
24. Tazi F, Ahsaini M, Khalouk A, Mellas S, Stuurman-Wieringa RE, Elfassi MJ, Farih MH (2012) Abscess of urachal remnants presenting with acute abdomen: a case series. Journal of medical case reports 6:226. https://doi.org/10.1186/1752-1947-6-226
25. Koster IM, Cleyndert P, Giard RW (2009) Best cases from the AFIP: urachal carcinoma. Radiographics: a review publication of the Radiological Society of North America, Inc 29 (3):939-942. https://doi.org/10.1148/rg.293085152
26. Hartman R, Kawashima A (2017) Lower tract neoplasm: Update of imaging evaluation. European Journal of Radiology 97:119–130. https://doi.org/10.1016/j.ejrad.2017.10.019
27. Wong-You-Cheong JJ, Woodward PJ, Manning MA, Davis CJ (2006) From the archives of the AFIP: Inflammatory and nonneoplastic bladder masses: radiologic-pathologic correlation. Radiographics: a review publication of the Radiological Society of North America, Inc 26 (6):1847-1868. https://doi.org/10.1148/rg.266065126
28. Dhillon J, Liang Y, Kamat AM, Siefker-Radtke A, Dinney CP, Czerniak B, Guo CC (2015) Urachal carcinoma: a pathologic and clinical study of 46 cases. Human pathology 46 (12):1808-1814. https://doi.org/10.1016/j.humpath.2015.07.021
29. Machida H, Ueno E, Nakazawa H, Fujimura M, Kihara T (2008) Computed tomographic appearance of urachal carcinoma associated with urachal diverticulum misdiagnosed by cystoscopy. Abdominal imaging 33 (3):363-366. https://doi.org/10.1007/s00261-007-9256-7
30. Willian Schmitt MB, Marco Ferreira, António Gomes, Ana Germano (2018) Urachal Adenocarcinoma: A Case Report with Key Imaging Findings and Radiologic-Pathologic Correlation. https://doi.org/10.1155/2018/4935261
31. Chen D, Li Y, Yu Z, Su Z, Ni L, Gui Y, Yang S, Shi B, Lai Y (2014) Investigating urachal carcinoma for more than 15 years. Oncology letters 8 (5):2279-2283. https://doi.org/10.3892/ol.2014.2502
32. Rafal RB, Markisz JA (1991) Urachal carcinoma: the role of magnetic resonance imaging. Urologic radiology 12 (4):184-187
33. Sheldon CA, Clayman RV, Gonzalez R, Williams RD, Fraley EE (1984) Malignant urachal lesions. J Urol 131 (1):1-8. https://doi.org/10.1016/s0022-5347(17)50167-6
34. Ashley RA, Inman BA, Sebo TJ, Leibovich BC, Blute ML, Kwon ED, Zincke H (2006) Urachal carcinoma: clinicopathologic features and long-term outcomes of an aggressive malignancy. Cancer 107 (4):712-720. https://doi.org/10.1002/cncr.22060
35. Pinthus JH, Haddad R, Trachtenberg J, Holowaty E, Bowler J, Herzenberg AM, Jewett M, Fleshner NE (2006) Population based survival data on urachal tumors. J Urol 175 (6):2042-2047; discussion 2047. https://doi.org/10.1016/s0022-5347(06)00263-1
36. Williams CR, Chavda K (2015) En Bloc Robot-assisted Laparoscopic Partial Cystectomy, Urachal Resection, and Pelvic Lymphadenectomy for Urachal Adenocarcinoma. Reviews in urology 17 (1):46-49
37. Aoun F, Peltier A, van Velthoven R (2015) Bladder sparing robot-assisted laparoscopic en bloc resection of urachus and umbilicus for urachal adenocarcinoma. J Robot Surg 9 (2):167-170. https://doi.org/10.1007/s11701-015-0507-2
38. Mertens LS, Behrendt MA, Mehta AM, Stokkel L, de Jong J, Boot H, Horenblas S, van der Heijden MS, Moonen LM, Aalbers AGJ, Meinhardt W, van Rhijn BWG (2019) Long-term survival after cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (HIPEC) for patients with peritoneal metastases of urachal cancer. Eur J Surg Oncol. https://doi.org/10.1016/j.ejso.2019.03.034
39. Mohile SG, Schleicher L, Petrylak DP (2008) Treatment of metastatic urachal carcinoma in an elderly woman. Nature clinical practice Oncology 5 (1):55-58. https://doi.org/10.1038/ncponc1009