PICTORIAL ESSAY | Urogenital imaging

Imaging evaluation of Urachus: Pictorial essay

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ABSTRACT

The urachus is a vestigial tubular structure extending from the bladder's anterior surface to the umbilicus, formed during embryonic development. Normally, it obliterates to form the median umbilical ligament; however, in some cases, remnants persist, leading to urachal anomalies such as fistulas, cysts, and diverticula. Urachal anomalies can result in complications, including infections and neoplasms, with malignant urachal adenocarcinoma being rare but more common than benign neoplasms. This cancer typically affects men aged 40-70, with symptoms like hematuria, abdominal pain, and palpable masses. Due to its extraperitoneal location, urachal carcinoma is often asymptomatic until advanced stages. Diagnosis of urachal carcinoma involves cystoscopy, imaging modalities like CT and MRI, and serum markers. Imaging plays a key role in detecting midline masses, often containing both solid and cystic components. CT and MRI can evaluate tumor extent and local invasion, but further assessment, such as biopsy, is often required. Surgical resection remains the primary treatment, with partial or radical cystectomy providing comparable outcomes. Despite advances in chemotherapy and immunotherapy, prognosis remains poor, with a 5-year survival rate of around 50%, particularly in cases with lymph node involvement or metastasis.

KEY WORDS: Urachus; Urachal cancer; Computed Tomography (CT); Magnetic Resonance Imaging (MRI)

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Introduction

The urachus is a tubular structure located in the retropubic space of Retzius between the transversalis fascia and the parietal peritoneum extending from the anterosuperior surface of the bladder to the umbilicus. [1,2] (Fig.1,2) It is formed from the involution of the embryological remnants' allantois and cloaca during the 4th month of pregnancy as the bladder descends into the pelvis [3]. Later on during pregnancy or shortly after birth it usually obliterates to form a cord-like structure called the median umbilical ligament. In some cases, this fails to happen and an urachus remnant is formed. There are four types of anomalies depending on the location of the abnormal residual potency along the tracheal tract: urachus or urachal fistula, urachal cyst (Fig.3a,b), umbilical-urachal sinus and vesicourachal diverticulum. [1] Patients with urachal anomalies can develop complications (Fig.4) including infection which is the most common one especially in children and neoplasms which usually occur in adult population. Benign neoplasms are extremely rare and have been described in the literature as case reports [1,3]. Malignant neoplasms are more common but still rare representing less than 1% of all bladder cancers [1].

Histology

Urachal remnants are lined with urothelium. However, and in contrast with bladder cancer (Fig.5) which is usually urothelial, urachus cancer is 80% adenocarcinoma more often mucin-producing or sometimes non-mucinous while sometimes a signet ring cell component may be present. The rest 20% of urachal malignancies are urothelial,sarcomatoid or squamous neoplasms [1,4]. It is believed that this is due to its chronic irritation and the consequent metaplasia of the transitional epithelium into columnar epithelium [1]. Furthermore, some adenocarcinomas may originate in embryonic rests of enteric cloacal cells [2,5]. Urachal adenocarcinoma represents 20-40% of all primary bladder adenocarcinomas [6].

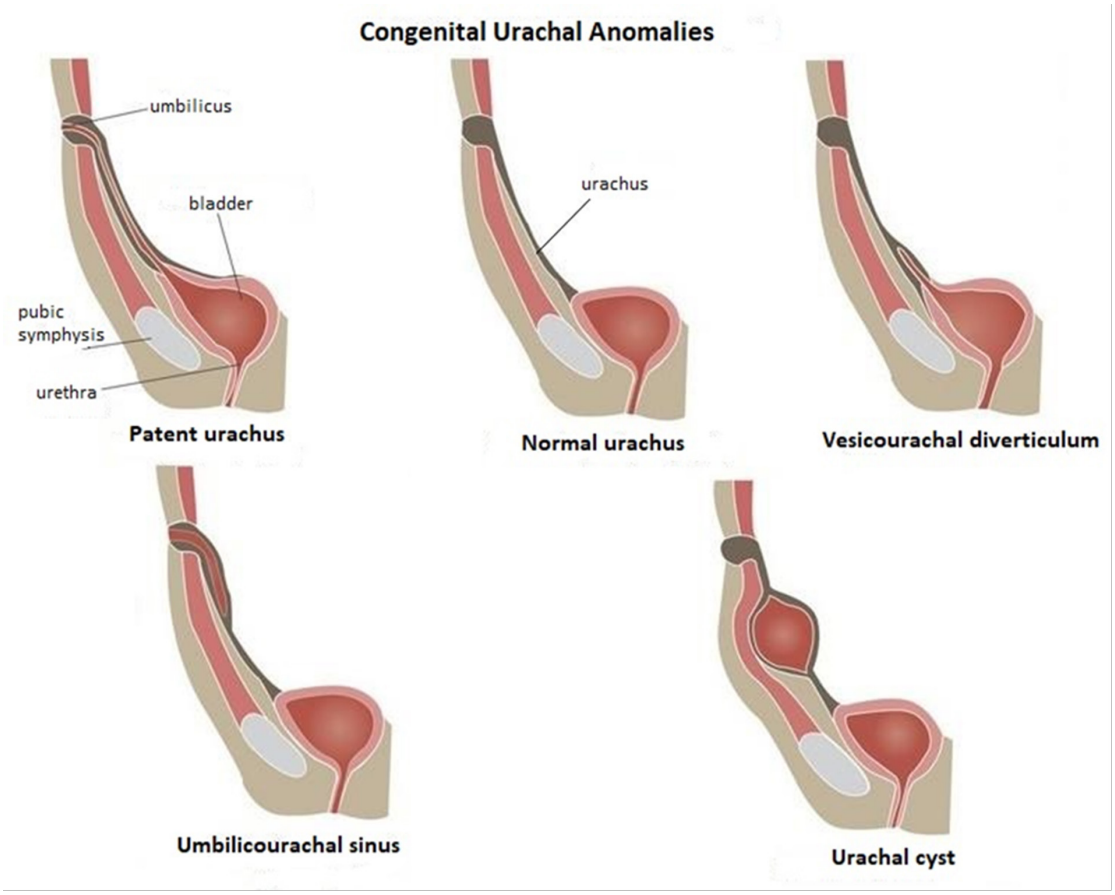
Clinical Findings and Diagnosis

Urachal carcinomas are considered clinically silent due to their location in the extraperitoneal space of Retzius and patients often present at advanced stages [7,3]. It more frequently affects male patients 40-70 years old. [2]. The most frequent symptom is macroscopic haematuria. Abdominal pain, dysuria, mucosuria and palpable lower abdominal mass can also be seen [8,9]. Nonspecific symptoms such as nausea, diarrhoea and weight loss are rare but they have also been reported [7].

Cystoscopy is a valuable diagnostic tool. Findings vary from bulging of the bladder wall from an external mass lesion with maintenance of normal mucosa at early stages to direct visualization of the tumor into the cavity of the bladder and estimation of possible ulceration at more advanced stages. [8,4] However, its accuracy rate in diagnosing urachal adenocarcinomas is considerably limited making it necessary to be combined with other diagnostic methods [8]. Serum markers such as carcinoembryonic antigen, carbohydrate antigen 19-9 and cancer antigen 125 are found increased in some cases due to the histopathological similarity of urachal adenocarcinoma with colorectal adenocarcinoma [4]. For the distinction between these two entities immunomarkers β-catenin and CK7 may be useful [7].

Imaging

Imaging plays a vital role in diagnosing urachal adenocarcinoma. Patients are often initially evaluated with ultrasound where it may appear as a heterogeneous midline soft tissue mass with increased vascularity [3]. At Computed Tomography (CT) urachal adenocarcinoma presents as a midline soft tissue mass most frequently with both cystic and solid components at the dome of the urinary bladder. It is adjacent to the lower abdominal wall and can extend into the space of Retzius. Calcifications may be seen in up to 70% of patients. They can be stipple, punctuate, curvilinear, psammomatous or peripheral. They are attributed to the mucinous component and they are considered nearly pathognomonic when found in a midline supravesical mass [7,3,6]. Urachal adenocarcinomas initially grow outside the bladder cavity and frequently extend to the adjacent bladder wall causing thickening and deformation [8]. The relationship between the tumor, bladder and urachus can be well demonstrated with thin slice sagittal reconstruction images [8].However CT has a limited value in evaluating tumor invasion of the bladder wall [4]. Magnetic Resonance Imaging (MRI)on the contrary can be helpful in better estimating the locoregional exten-



(Fig.1) *Figures of the normal urachus and the types of congenital anomalies.*

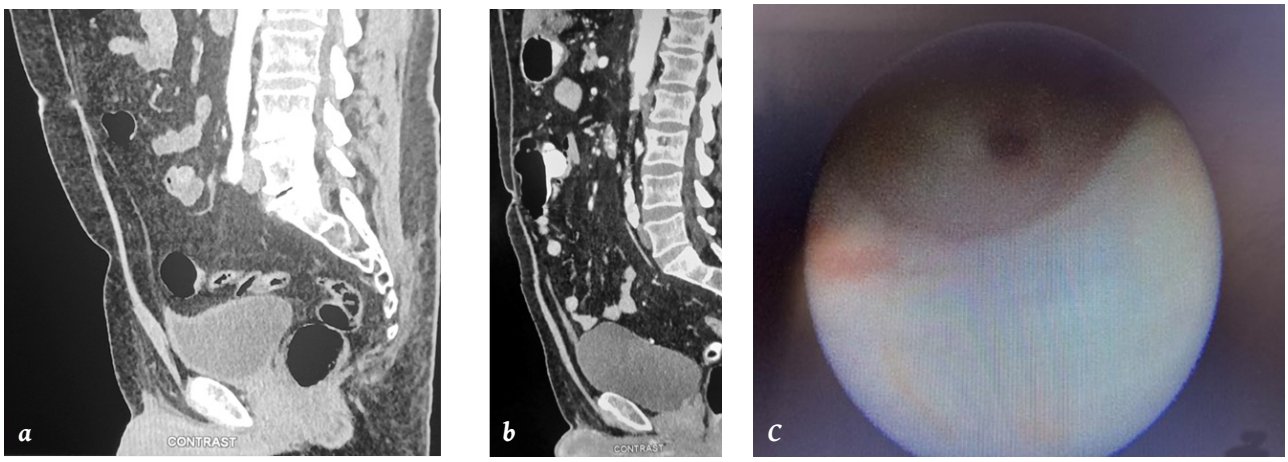
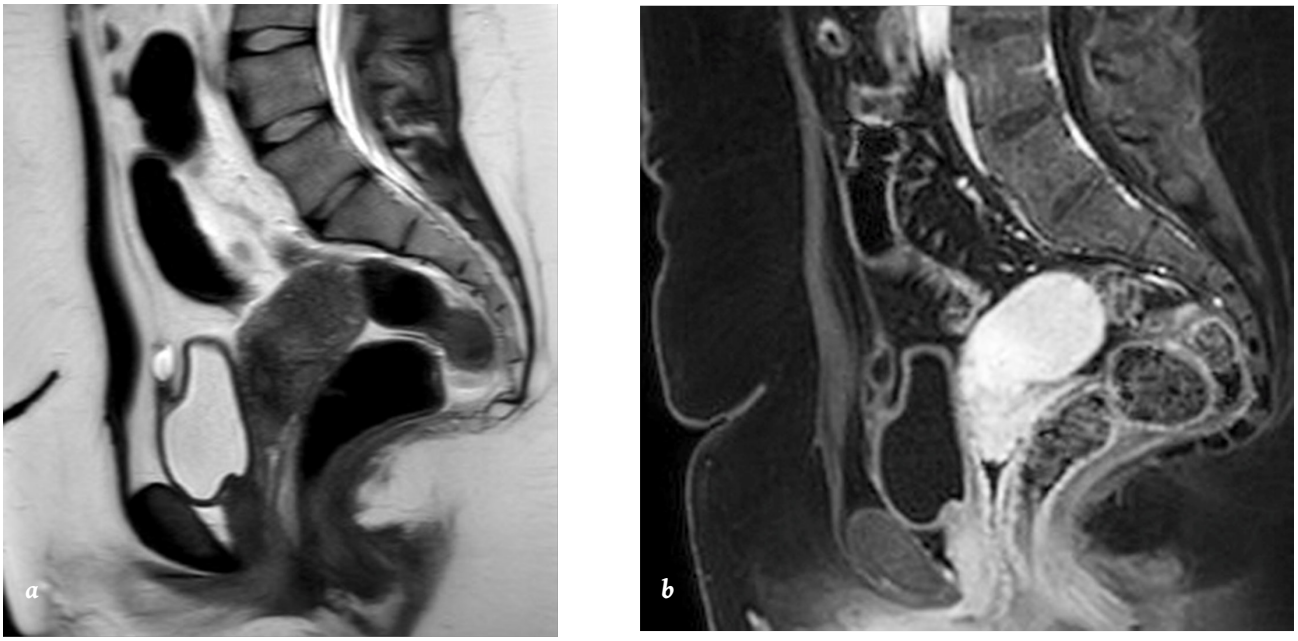


Fig.2a,b *Normal urachus on CT sagittal reconstructions*
(Fig. 2c) *Cystoscopic view of the opening of the urachus*



(Fig.3a,b) MRI: Urachal cyst

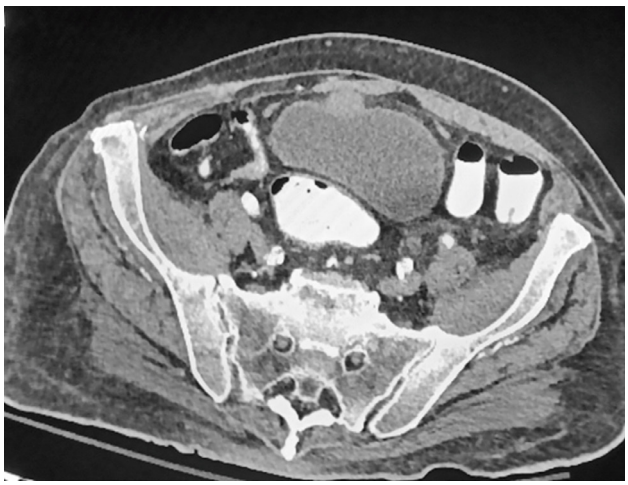
sion of the lesion. Urachal adenocarcinomas show variable signal intensity on T2 which tends to increase as the amount of mucus into the lesion increases [6]. High T2 signal may also be due to fluid collection or necrosis but in that case the post contrast enhancing glandular tissue is missing compared to high T2 signal due to mucinous elements [1]. Solid components are isointense to soft tissue on T1-weighted images and they enhance heterogeneously after contrast administration [1,3]. Diffusion- weighted MRI sequences play a useful role in differentiating malignant from non-malignant urachal anomalies but their sensitivity is still relatively low and further investigation with fine needle aspiration and/or cystoscopy with biopsy may be necessary for further investigation [5]. CT or MR can also provide useful information regarding pelvic lymph node involvement and distant metastasis which occurs in later stages and usually involves the lung, liver, omentum, iliac/inguinal lymph nodes and bone [2]. Although FDG-PET CT plays a decisive role in staging malignancies its use is limited for urachal adenocarcinomas which are relatively hypocellular due to their often mucinous origin and therefore demonstrate low or even absent FDG uptake [6].

Differential Diagnosis

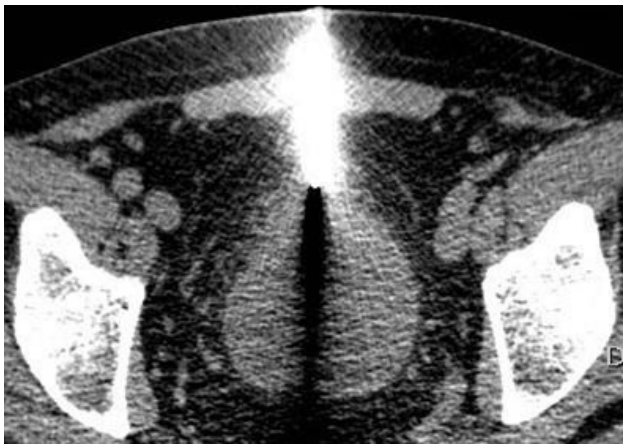
Although the list of Differential Diagnosis is not long the distinction between the entities included may be impossible with the available diagnosing methods and surgical treatment is often required to exclude malignancy. Non-urachal carcinoma of the bladder mostly located in the later wall, can be multiple and show early clinical symptoms. Other neoplasms included in the differential diagnosis are benign urachal tumors such as adenomas, fibromas, hamartomas, etc) which are considered extremely rare and metastasis from other malignancies [6]. Infection of urachal remnant and formation of abscess can also mimic cancer. Both entities show similar appearance on both CT and MR but there are several elements such as the presence of calcifications, bladder wall invasion or distant metastasis that indicate malignancy [5]. Diffusion weighted MRI sequences also contribute to the emergence of malignancy [5].

Managment and Survival

Surgery is the gold standard for treating non-metastatic urachal carcinoma. Partial and radical cystectomy are both considered reasonable and provides similar oncological outcomes. One of the most significant risk factors are positive surgical margins and therefore en-block resection and removal of urachal remnant and the umbilicus is necessary [4]. The therapeutic role of pelvic lymphadenectomy is controversial. Chemotherapy may

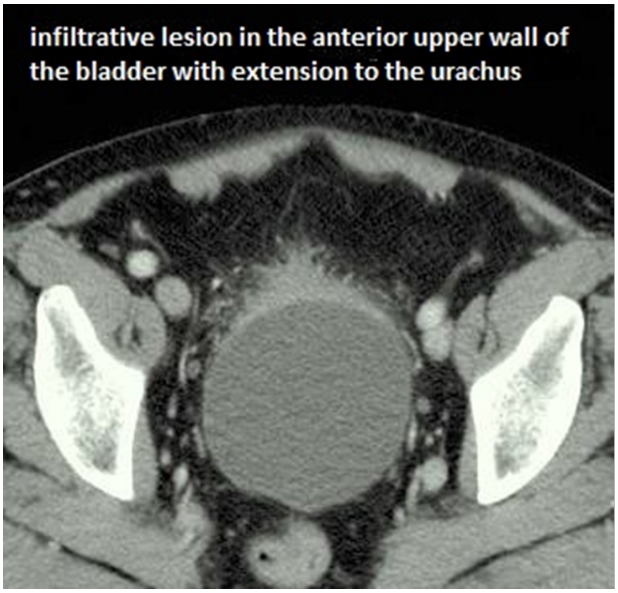


(Fig.4) *CT: Inflammation of urachus*



(Fig. 5b) *CT-guided core biopsy of the tumor*

also be used either as neoadjuvant in cases of unresectable tumors at presentation or when metastasis is present. The most common regimens are 5-fluorouracil and cisplatin or FOLFOX6 (folinic acid, 5-fluorouracil and oxaliplatin) [9]. Immune targeting therapies although promising need further investigation with clinical studies to be well established [9]. Prognosis for urachal carcinoma is considered poor and the 5-year overall survival rate is found to be ~50% [9]. The presence of lymph nodes, distant metastasis or positive surgical margins are considered poor prognostic factors [4].



(Fig.5a) *CT: Tumor of the bladder with extension to the urachus*

Conclusion

Imaging plays an essential role in the evaluation of urachal anomalies and along with other diagnostic tools can contribute to the correct diagnosis and management of such patients. **R**

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