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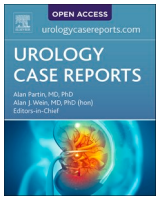

Oncology

# Rare case of enteric type of urachal adenocarcinoma: A case report

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ABSTRACT

Urachal adenocarcinomas, constituting 10 % of bladder adenocarcinomas, pose a significant challenge with limited literature. A 43-year-old male presented with haematuria and abdominal pain, leading to surgical intervention for a 13 cm pelvic tumor. Histopathology identified an intestinal-type primary urachal adenocarcinoma, staged as IIIA, no recurrence on follow-up. Early detection is crucial for improved outcomes in these rare malignancies. While surgery remains the primary treatment, outcomes vary, emphasizing the need for research on standardized protocols. Enhanced awareness and interdisciplinary collaboration are vital for effective management. Comprehensive guidelines are essential for optimizing patient prognoses in urachal adenocarcinomas.

## 1. Introduction

Adenocarcinomas arising within the urinary bladder present a notable challenge in the landscape of urological malignancies, constituting a minority subset, with an incidence ranging between 0.5 % and 2.0 % of all bladder carcinomas. Among this already rare classification, urachal adenocarcinomas emerge as a distinctive entity, accounting for approximately 10 % of cases or approximately one-third of all bladder adenocarcinomas. The urachus, an embryonic structure that connects the bladder to the umbilicus, can undergo carcinomatous changes, but this is rare.[1,2]

Although urachal adenocarcinoma is clinically significant, the literature is notably sparse, with a paucity of comprehensive studies and a reliance on retrospective analyses and sporadic case reports. Such scarcity in documented cases has contributed to challenges in understanding the disease's diverse clinical presentations, optimal diagnostic strategies, and tailored treatment approaches.

In this context, we present a case report delineating the clinical course of a male patient diagnosed with urachal adenocarcinoma, notable for its unusual manifestation in the absence of significant past medical history. This case illuminates the complexities associated with diagnosis, management, and treatment decisions in an exceedingly rare presentation of urachal adenocarcinoma.

The objective of this report is not only to contribute a singular clinical instance but also to augment the existing knowledge base, providing clinicians with a reference point to heighten vigilance regarding this infrequently encountered pathology and to facilitate more timely and accurate diagnosis and management.

Through a detailed exposition of this case, we aim to provide a comprehensive understanding of the clinical nuances, challenges, and considerations associated with urachal adenocarcinoma, thereby advocating for improved awareness and enhanced strategies for its effective clinical management.

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## 2. Case report

A 43-year-old man complained of haematuria and spastic abdominal pain for two years to a general practitioner. Ignoring the symptoms and attributing them to kidney stone disease until a severe lumbar pain attack that had lasted for about 24 hours when he decided to search for medical help. An ultrasonography of the abdomen detected a cystic abdominal mass. Later he was hospitalized in the "Second Surgery" department of UMHAT „St. George" Plovdiv, Bulgaria where a second abdominal ultrasonography confirmed a 13 cm large pelvic tumor. A contrast-enchanced CT revealed a tumor formation originating from the upper-anterior wall of the urinary bladder and growing cranio-medially to the abdominal cavity, right below the anterior abdominal wall. The shape of the formation was oval, elongated, and with sharp contours. The dimensions were: longitudinal - 129 mm, transverse - 67 mm, antero-posterior - 66 mm. [Fig. 1]. The formation was with a cystic character, shell-like calcifications on the periphery were present, a calcium-dense structure in the interior was described and a solid nodular part, absorbing the contrast substance located in the region of the caudal pole. The density of the cyst fluid was about 25 HU, remaining unchanged to contrast. The surrounding adipose tissue was intact. As an additional finding a paravesical lymph node on the right with a size of 7.5 mm, a para-aortic lymph node −7.5 mm, several mesenteric lymph nodes with sizes up to 6 mm and pyelonehritis of the left kidney were described.

No tumor markers were tested preoperatively. Laboratory findings were within the normal ranges except slightly low level of hemoglobin (135g/l), increased levels of platelet cells (486 10^9/l), red blood cells (6.26 10^12/L) and cholesterol (6.8 mmol/L).

A team of general surgeons and an urologist was built up and a laparotomy was performed. During the procedure, a sizable cystic mass that stretched from the umbilicus to the bladder dome was detected. The bladder examination revealed an absence of tumor, but the cystic formation appeared to be a straight extension of the dome. Radical removal of the tumor with partial resection of the urinary bladder, urachal and umbilical resection including para-aortic lymph node dissection were performed.

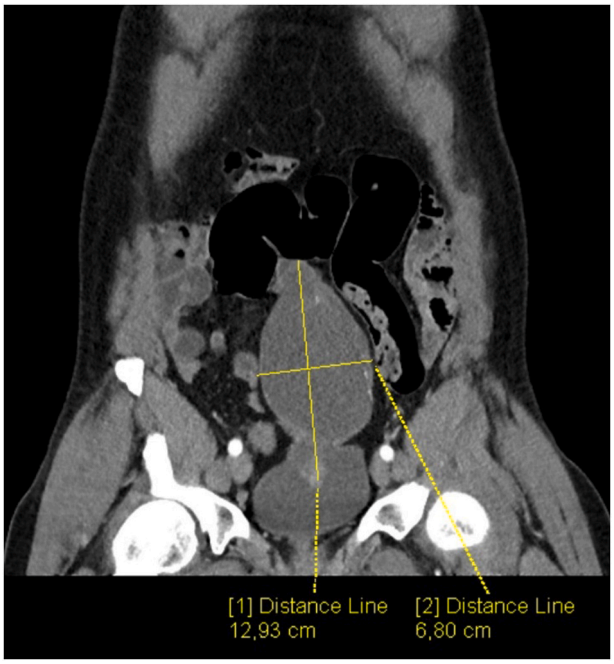


**Fig. 1.** Coronal CT image depicting a large and heterogeneous median abdominal mass, extending from the anterosuperior aspect of the bladder towards the umbilicus.

The frozen section procedure revealed intestinal type of primary urachal adenocarcinoma. Immunohistochemistry analysis showed a strong immunoreactivity for CK20, CDX2 and high molecular weight keratin (HMWCK) and the histopathological diagnosis established was intestinal type of primary urachal adenocarcinoma without lymphatic metastatic spread [Fig. 2].

The tumor diameter was 13 cm,.There was no evidence of invasion in the surrounding areas. The surgical margins were found to be free of any tumor cells. The patient's condition was assessed as stage IIIA according on the Sheldon 1984 classification, and a positron emission tomography/computed tomography performed a month following the operation showed no local recurrence, no lymphatic or distant metastases [Fig. 3].

The patient did not get any additional treatment, with a planned follow-up period of five years after three months of monitoring, there are no reported symptoms or signs of disease. Informed consent was obtained from the patient.

## 3. Discussion

The urachus is a residual structure from the urogenital sinus and allantois during embryonic development. It has an average length of 5–5.5 cm and is positioned between the transversalis fascia in the front and the parietal peritoneal layer in the back. Later in pregnancy, it spontaneously involutes into the median umbilical ligament, a fibrous cord[1–4] Partial involution might be observed in approximately 32 % of adults and although rarely, might be associated with various neoplasms the vast majority of which of epithelial origin. The remaining epithelium is typically composed of urothelial cells, however the majority of urachal carcinomas are of glandular type. It is hypothesized that these carcinomas develop from intestinal metaplasia of the remaining lining or from the malignant transformation of persistent ectopic intestinal tissue originating from the cloaca.[5] Hue and Jacquin were first to document the presence of urachal carcinoma in 1863. Later, in the 1930s, C. Begg further advanced our knowledge of urachal neoplasms, establishing the basis for our current understanding.[5]

Carcinoma of the urachus is a rare malignancy accounting for 0.2 % of all bladder cancers and affecting mainly the age range of 45–75 year-old with a male-to-female ratio of 1.4–1.[6] Urachal adenocarcinomas often occur in a midline position involving the dome of the bladder and retropubic space.[7] In most of the cases it is located at the junction of the lower urachus and the urinary bladder.[8]

Urachal carcinoma encompasses various histologic subtypes, with adenocarcinomas being the most prevalent (>90 %).[9–12] Adenocarcinomas can be categorized into two distinct groups: cystic and noncystic adenocarcinomas.[13] The majority (83 %) of urachal carcinomas are noncystic,[14] with mucinous being the most prevalent subtype (50 %),

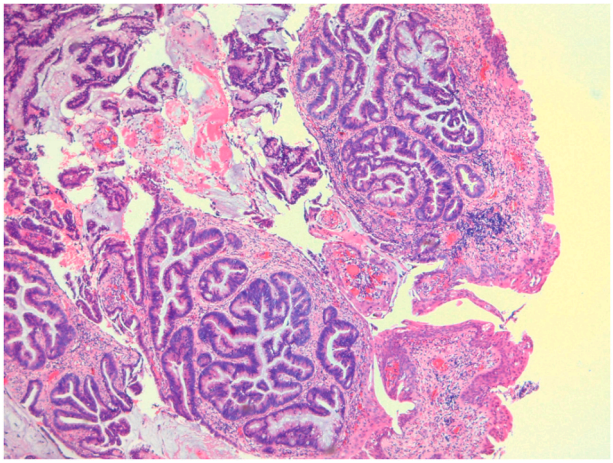

**Fig. 2.** Histopathological material of enteric type urachal adenocarcinoma.

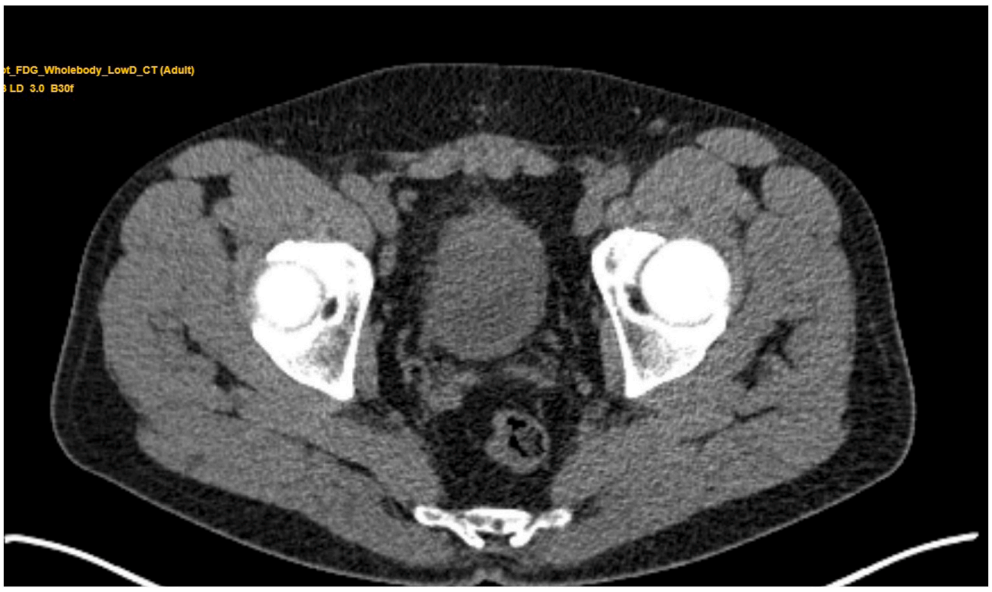


**Fig. 3.** CT image showing no local recurrence or lymphatic metastases.

followed by enteric (24 %), mixed (10 %), not otherwise defined (9 %), and signet ring cell (7 %).[10,14–17] Urachal adenocarcinomas can also contain a small amount of non-glandular carcinoma, which is observed in 4 %–8 % of cases.[10,14]Around 70 % of urachus adenocarcinomas exhibit the secretion of mucin and the formation of calcifications. This histological feature indicates that urachus adenocarcinomas originate from the glandular intestinal epithelium, which is generated as a result of the presence of cloaca residue or enteric inclusion. A study has confirmed the presence of monoclonal antibody against the colonic epithelial protein in samples of urachal carcinoma, thus substantiating this concept.[18] Thus, urachal adenocarcinomas seem to be a distinct entity on the molecular level with closer resemblance to colorectal adenocarcinomas than to urothelial carcinomas.[12] Sarcomas, small-cell carcinomas, transitional epithelial cell, and mixed neoplasms are rarely seen.[18]

About 8 % of patients are asymptomatic,[15] and when symptomatic urachal adenocarcinomas present with haematuria, mucinuria, palpable mass, bacteriuria, umbilical or pelvic pain and weight loss.[9,14,15,19] Diagnosing urachal carcinoma accurately is difficult due to its histologic similarities with other adenocarcinomas and its close proximity to the bladder and colorectum. It necessitates a careful clinico-pathologic correlation and adherence to specific diagnostic criteria in order to establish a definitive diagnosis.[5] Standard imaging workup for urachal cancer includes ultrasonography, CT scan, and/or MRI evaluation of the abdomen and pelvis.[20] Immunohistochemical characteristics are employed to aid in the diagnosis of conditions that are challenging to differentiate. Urachal adenocarcinoma is frequently characterized by the presence of Cytokeratin 7 and CD15 (LeuM1), which aid in distinguishing it from colorectal adenocarcinoma. The β-catenin marker typically exhibits nuclear positive in colorectal adenocarcinoma, while this is less frequently observed in urachal adenocarcinoma.[17] CEA and CA19-9 levels increase in these patients due to their histopathological features similar to the enteric epithelium.[19] Elevated serum CA 125 levels can also be observed in certain patients with abdominal carcinomatosis.[20] The prognosis for individuals with urachal adenocarcinoma is typically unfavorable due to the tumor's location, delayed symptom development, and high likelihood of metastasis. The prognosis is mostly influenced by the grade of the tumor and the adequacy of surgical margins.[11]

In 1984, Sheldon[11] devised a taxonomy for urachal tumours. [Table 1].

Mayo Clinic has devised a novel and streamlined stage method. [Table 2].

**Table 1**
Sheldon staging system for urachal carcinoma.

| | |
|---|---|
| Stage I: No invasion beyond the urachal mucosa | |
| Stage II: Invasion confined to the urachus | |
| Stage III: Local extension into the: | Bladder (IIIA) |
| | Abdominal wall (IIIB) |
| | Peritoneum (IIIC) |
| | Viscera other than bladder (IIID) |
| Stage IV: Metastases to the: | Regional lymph nodes (IVA) |
| | Distant sites (IVB) |

**Table 2**
Mayo clinic staging system for urachal carcinoma.

| | |
|---|---|
| Stage I | Tumours confined to the urachus and/or bladder |
| Stage II | Tumours extending beyond the muscular layer of the urachus and/or the bladder |
| Stage III | Tumours infiltrating the regional lymph nodes |
| Stage IV | Tumours infiltrating non regional lymph nodes or other distant sites |

Currently, both staging systems are utilized.[19]

Urachal cancer being one of the rarest malignancies of the urinary bladder and having late clinical manifestations is usally diagnosed at an advanced stage with life expectancy not being more than a year for up to 60 % of stage IV tumours.[19] The mean survival for a locally advanced or metastatic disease is between 12 and 24 months, and the 5-year survival rate is only 43 %.[20] The areas where local recurrence commonly occurs include the pelvic lymph nodes, peritoneum, and omentum. The sites where distant metastasis occurs include the lungs, lymph nodes, bones, intestines, brain, and liver.[19] Metastatic disease is commonly found at the time of diagnosis in as many as 1 in 5 patients.[20]

Wide local excision of urachal mass with umbilicus and surrounding soft tissue en bloc combined with partial or radical cystectomy and bilateral pelvic lymphadenectomy is considered to be the primary surgical management. However, many publications in literature report that en bloc removal of tumor with umbilicus, entire urachal ligament, and bladder dome alone has long-term survival and disease-free period.[19] To guarantee complete removal of the urachal ligament and umbilicus, it is

advisable to perform further resection, as there is a 7 % chance of urachal cancer developing at the umbilicus.[20] Recurrence risk factors after surgery include the presence of positive margins, lymph node involvement, or additional metastases during surgery, as well as the failure to remove the umbilicus.[18] Currently, there is a lack of established guidelines for the treatment of urachal adenocarcinoma with adjuvant chemotherapy. The efficacy and advantages of chemotherapy and radiation therapy in patient treatment remain uncertain.[20]

## 4. Conclusions

Urachal adenocarcinomas are rare neoplasms usually presenting as locally advanced tumours or with distant metastatic spread at the time of diagnosis. One of the most common symptoms is haematuria. The primary treatment option is surgery which in cases with an advanced disease may also include radio- and chemotherapy. The prognosis varies, but timely consultation with a healthcare professional and early detection and intervention generally improve outcomes.

## CRediT authorship contribution statement

**Petar Uchikov:** Writing – original draft, Data curation. **Nedzhat Ali:** Writing – review & editing, Conceptualization. **Krasimir Kraev:** Writing – original draft, Data curation. **Bozhidar Hristov:** Visualization, Resources. **Atanas Ivanov:** Resources, Formal analysis. **Mladen Doykov:** Writing – review & editing, Formal analysis. **Maria Koleva-Ivanova:** Project administration, Methodology. **Angelina Mollova-Kyosebekirova:** Visualization, Validation. **Maria Kraeva:** Resources, Data curation. **Dzhevdet Chakarov:** Validation, Supervision. **Milena Sandeva:** Formal analysis, Conceptualization. **Bistra Dobreva-Yatseva:** Data curation, Supervision. **Petar Antonov:** Investigation, Conceptualization.

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