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## Case report

# Primary urachal adenocarcinoma treated effectively with surgery and post operative chemoradiation therapy: Case Report with review of the literature <sup>☆</sup>

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## ABSTRACT

Urachal carcinomas is a rare and aggressive tumor, accounting for less than 1% of all bladder cancers. We report a case of a 32-year-old man, with no past medical history, complaining of a total hematuria. The abdominal computed tomography scan revealed an exophytic mass of 3 cm on the dome of the bladder, extending to the urachus. The computed tomography scan of chest, abdomen and pelvis did not show neither regional or distant metastasis. Partial Cystectomy with umbilectomy was performed. Histopathology was in favor of urachal adenocarcinoma, classed pT3a, based on Sheldon's staging system, pT2b based on Mayo system, and pT2 based on Ontario system.

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### Learning objectives:

- Urachal carcinomas is a rare and aggressive tumor, accounting for less than 1% of all bladder cancers.
- Adenocarcinomas is the most frequent histologic subtype.

- The standard Imaging examinations include abdominal and pelvic ultrasonography, computed tomography and computed tomography chest to rule out lung metastases.

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- Surgery is the most effective treatment for resectable lesions. It includes a partial cystectomy with Umbilectomy.
- Adjuvant radiotherapy improves progression free survival in case of close or positive margins or an unresectable lesion.

## Introduction

The urachus is a vestigial structure that connects the bladder to the allantois during early embryonic development. At 32 weeks of pregnancy, the urachus involutes and became a fibrous cord known as the median umbilical ligament. If remnants of the allantois remains within the ligament, they may turn into cysts and epithelial neoplasms.

Urachal carcinomas is a rare and aggressive tumor, accounting for less than 1% of all bladder cancers [1], Hue and Jacquin published the first case report in 1863 [2], followed by an elaborate work by T. Cullen in 1916 [3]. Sheldon et al proposed the initially accepted a staging system for the urachus carcinoma revised by the Mayo Clinic's team in 2006 [4,5].

Histologically, adenocarcinoma is an uncommon histologic variant and accounts for 1.4 % of bladder cancers. Among these rare cases of adenocarcinoma of the bladder, urachal cancer represents 10%-40%.

To date, and because of its rarity, knowledge of the patient demographics, the diagnostic criterias, the staging system to use, the management and survival of this uncommon malignancy is poorly known.

Urachal carcinoma occurs at a relatively younger age than bladder carcinoma, and given the silent nature of early lesions, it often presents at an advanced stage and has a poor prognosis.

We present in this paper a case of urachal adenocarcinoma in a 32-year-old man, with advanced stage disease. That we treated with partial cystectomy in addition to adjuvant radiotherapy, based on a review of the literature.

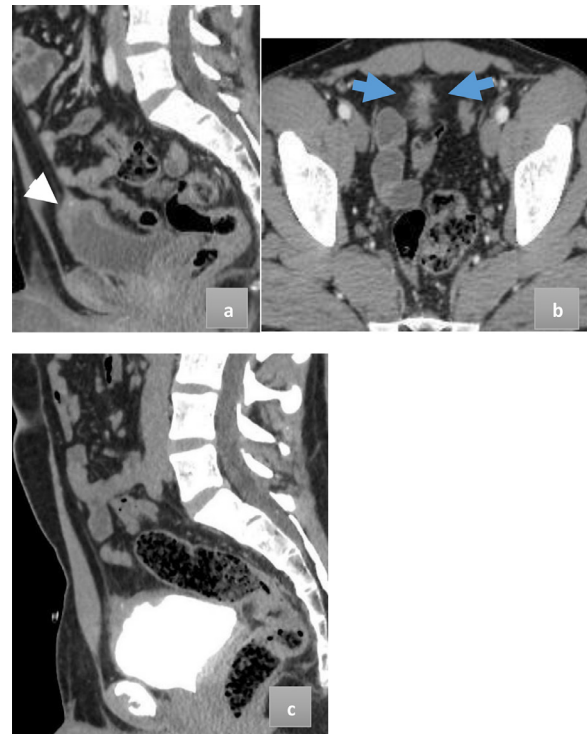
## Case report

32-year-old man with no remarkable past medical and family history, who consulted a urologist because of painless macroscopic hematuria for the last four months.

Otherwise, he had neither specific symptoms nor a loss of body weight. There were no abnormalities in the physical examination and on biochemical parameters.

The cystoscopy evaluation showed a solid tumor, which measured about 3 cm in the bladder dome. The lesion was resected and the histology revealed infiltration of the bladder mucosa by mucinous adenocarcinoma.

The computerized tomography scan analysis confirmed the presence of an intra-abdominal mass with calcification and adjacent bladder dome invasion with no communication with urachus. (Fig. 1). No regional nor distant metastasis were shown by The CT scan of chest-abdomen and pelvis.



**Fig. 1 – sagittal (A, C) and axial (B) contrast-enhanced Uro-CT images, shows a midline irregular thickening of the wall of bladder dome with some calcifications (head of arrow) with invasion of the perivesical fat (blue arrows). There is a communication with urachus. (Color version of figure is available online).**

A partial cystectomy of the dome was performed with total urachectomy up to the umbilicus and bilateral pelvic node dissection. Histo-pathological examination of the specimen revealed infiltration of the bladder muscle by mucinous urachal adenocarcinoma pT3a based on Sheldon's staging system, pT2b based on Mayo system and pT2 based on Ontario system. No adjuvant therapy was considered since the surgical margins and nodes were negative.

A CT scan performed 6 months after the surgery, confirmed local recurrence and a total cystectomy was planned. The patient refused the surgical revision and he was referred to the radiotherapy department for further management. We opted for a concomitant chemoradiotherapy with radiation dose of 66 Gy (2Gy/fraction) associated with 3 cycles of 5FU and cisplatin chemotherapy regimen.

Two years after radiation, there were no signs of primary disease or metastases in other organs.

## Discussion

Urachal carcinomas is a rare and aggressive tumor, accounting for less than 1% of all bladder cancers [6]. It is a quite rare entity, and most of the reported studies are single-cases or small-series.

Because of the silent nature of early lesions, the disease is usually advanced when the symptoms appear. The common metastatic sites include the lymph nodes, peritoneum and lung.

The used modalities for the initial diagnostic may include cytology, imaging and cystoscopy. The Cystoscopy can identify a visible mass in about 80% of patients, whereas urine cytology will be positive in only 38%. Ultrasound scan is often the primary imaging modality, the tumor is observed as a soft tissue mass, with some heterogeneities and calcification. On CT scan, Urachal cancer usually appears as an intra-abdominal mass adjacent to the bladder dome, extending into the space of Retzius. It can be cystic, solid, or mixed. Additionally, calcification within the tumor occurs in 50%-70% of cases which is considered highly suspicious, if not pathognomonic, for urachal cancer. Magnetic resonance imaging is also helpful, although usually performed for evaluation of unrelated disease, with the cyst diagnosed as an incidental finding. The sagittal images are important to define the location of the tumor.

The use of The TNM staging system for urachal adenocarcinoma is limited as some urachal adenocarcinomas do not invade the bladder's surface urothelium. Currently, several specific staging systems for urachal tumors have been used but neither system has been validated. Sheldon et al [7] proposed a staging system in which stage I and II tumors were defined on the basis of urachal confinement, while late stage disease involves the local structures, such as the bladder, abdominal wall or peritoneum, and metastases to regional lymph nodes or distant sites. In 2006, a more simplified system has been proposed by Ashley et al. [5] at the Mayo Clinic the Mayo clinic, 4 stages were defined as so: Stage I, tumors confined to the urachus and/or bladder; Stage II, tumors extending beyond the muscular layer of the urachus and/or the bladder; Stage III, tumors infiltrating the regional lymph nodes; and Stage IV, tumors infiltrating non regional lymph nodes or other distant sites. The Ontario staging system is yet another simplified classification of urachal tumor involving 4 stages: confined to urachus (T1), confined to bladder (T2), Invading surrounding fat (T3), and extending to the peritoneum (T4) [8].

Bladder cancer is the most common malignancy involving the urinary system. Urothelial carcinoma is the predominant histologic type in the United States and Europe, in over 90% of cases. Primary adenocarcinoma of the urinary bladder can be classified as urachal adenocarcinoma and non urachal adenocarcinoma. It is often difficult to differentiate between the two entities because of the overlapping histologic and immunohistochemical features.

Several diagnostic criteria have been proposed for the diagnosis of urachal adenocarcinoma. Wheeler et al [9] initially proposed a set of strict criteria, which included the following:

(1) tumor in the dome of the bladder, (2) absence of cystitis cystica and cystitis glandularis, (3) predominant involvement of the muscularis or deeper tissue by tumor with intact or ulcerated epithelium, (4) presence of urachal remnants, (5) presence of a suprapubic mass, (6) sharp demarcation between the tumor and the normal surface epithelium, and (7) tumor growth in the bladder wall, extending into the space of Retzius.

However, these criteria were too restrictive and very few reported cases of urachal carcinoma would meet all the requirements.

Subsequently, Johnson et al. modified the criteria and included the following:

(1) tumor in the dome or elsewhere in the midline of the bladder, (2) sharp demarcation between the tumor and the surface epithelium, and (3) absence of primary adenocarcinoma of another organ. This classification is practical and has been widely adopted.

Recently, The MD Anderson Cancer Center (MDACC) suggested 5 criterias for the diagnosis of urachal cancers [10]. These criteria include a mid-line location of the tumor; a sharp demarcation between the tumor and normal surface epithelium; an enteric histology; the absence of urothelial dysplasia, cystitis cystica or cystitis glandularis transitioning to the tumor; and the absence of a primary adenocarcinoma of another origin.

The therapeutic strategy for urachal carcinoma has not yet been established because of its rare occurrence. Management strategies hinge upon partial cystectomy with large resection of the urachus and umbilicus, as the tumor predominantly involves the muscularis propria and perivesical soft tissue and the bladder surface urothelium is usually not affected. In the Mayo clinic series, the authors concluded that complete urachectomy and umbilectomy were significant predictors of survival on univariate analysis, and recommended their routine inclusion.

Urachal tumors are not particularly radiosensitive and radiotherapy is rarely used. Radiation therapy can be considered for postoperative positive margins and for localized inoperable disease, but the benefits are unclear. Prognostic is often poor due to the advanced stage of the disease at diagnosis.

A recent retrospective review of urachal carcinomas diagnosed from 1994-2011 from the National Cancer Registry in Ireland involved a total of 26 histo-pathological confirmed urachal carcinomas [11].

Authors reported a median overall survival of 2.7 years following diagnosis (range 0-15.2 years).

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## Conclusion

Urachal cancer is a rare and aggressive tumor with a poor prognosis when presenting with advanced stage. Despite limited evidence, partial cystectomy with wide resection of the urachus and umbilicus is the gold standard in localized cases.

Further investigation is warranted to define the best therapeutic strategy to improve recurrence-free and overall survival rates.

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## Patient consent

Patient consent statement A written consent was obtained from the patient for publication of this case.

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