Section: General Surgery

Case Report

FROM CRIMSON CLUES TO THE UNSEEN ENEMY: NAVIGATING THE THERAPEUTIC MAZE OF URACHAL CARCINOMA – A COMPELLING CASE REPORT

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Abstract

Background: Urachal carcinoma is a rare, aggressive form of bladder cancer, representing only 0.01% of all adult malignancies. Due to its embryonic origin from the allantois, it often presents late with nonspecific symptoms, complicating timely diagnosis and treatment. This case report outlines the clinical presentation, diagnosis, surgical management, and histopathological findings of a high-grade urachal adenocarcinoma. **Case Presentation:** A 64-year-old male presented with lower abdominal pain, hematuria, and increased frequency of micturition. Examination revealed a firm, non-tender midline mass in the suprapubic region. Imaging suggestive of mass infiltrating the bladder dome with polypoid growth into the lumen and abutting the anterior abdominal wall. Cystoscopy revealed a large proliferative growth, and biopsy indicated high-grade noninvasive papillary urothelial carcinoma. The patient underwent umbilicectomy, urachal cyst excision, partial cystectomy, and bilateral pelvic lymph node dissection. Histopathology confirmed adenocarcinoma of urachal origin characterized by glandular structures with mucin production. **Conclusion:** Early recognition of urachal carcinoma symptoms, combined with imaging and histological confirmation, is critical for appropriate surgical intervention. Comprehensive resection remains the cornerstone of treatment, with ongoing research needed for adjuvant therapies to improve long-term survival.

INTRODUCTION

Urachal carcinoma is a rare and aggressive type of bladder cancer, accounting for only about 0.01% of all adult malignancies. Originating from remnants of the embryonic allantois, it often presents at an advanced stage with vague, nonspecific symptoms, making early diagnosis and treatment challenging. This case report describes the clinical presentation, diagnostic workup, surgical management, and histopathological characteristics of a high-grade urachal adenocarcinoma.

CASE REPORT

A 64-year-old male who is a known hypertensive, presented with complaints of lower abdominal pain, hematuria and increased frequency of micturition. On physical examination, an ill defined firm, and non tender mass was palpable, in the midline of suprapubic region.

Initially ultrasound abdomen was done, revealed a 6x7cm solid cystic lesion in relation to superior wall of urinary bladder. Subsequent contrast CT showed a well defined solid cystic lesion abutting the dome of urinary bladder towards the midline, 6.6x4.5x7.4cm in size, solid component showed contrast enhancement. Lesion seen infiltrating the urinary bladder with polypoidal growth into the bladder lumen. Anteriorly, lesion is abutting the abdominal wall with maintained fat plane.

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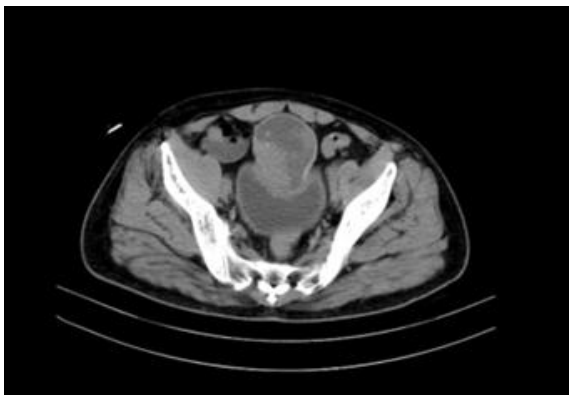

Figure 1: Axial image, contrast-enhanced CT scan of the abdomen and/or pelvis demonstrates well defined solid cystic lesion abutting the dome of urinary bladder.lesion seen infiltrating the urinary bladder with polypoidal growth into the lumen, anteriorly lesion abutting abdominal wall with maintained fat plane

Following which, cystoscope was done, detected a large proliferative growth over the anterior wall of bladder, towards the apex. Biopsy was taken and the result came as Noninvasive papillary urothelial carcinoma high grade. The patient underwent Umbilectomy, urachal cyst excision, partial cystectomy and bilateral pelvic lymph node dissection.

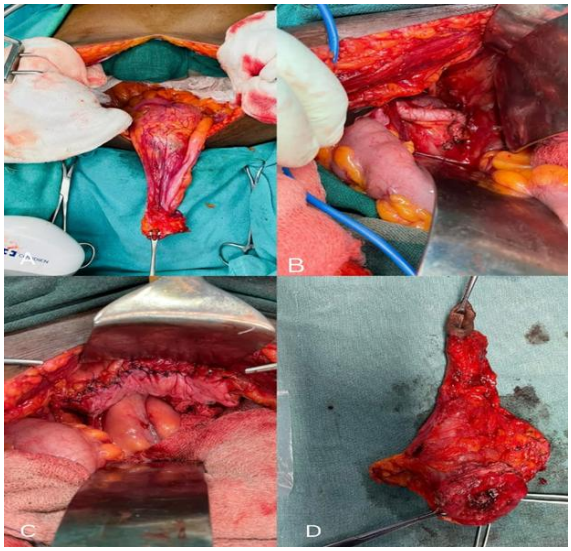

Figure 2: A. Intraoperative picture showing the tumor, B.Intraoperative view after pelvic lymph node dissection, C.Intraoperative appearance of residual bladder after partial cystectomy, D. Resected urachal tumor with umbilicectomy and partial cystectomy

Histopathology showed adenocarcinoma of urachal origin NOS.pT3A

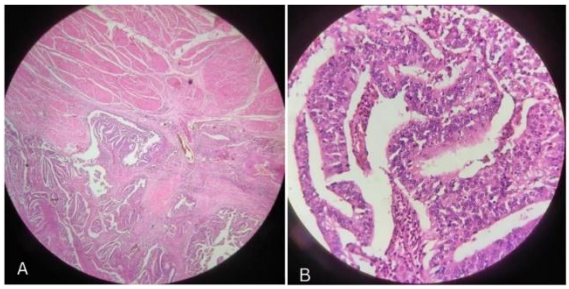

Figure 3. Histopathological picture showing infiltrating neoplasm composed of cells arranged in glandular pattern (A.Low resolution B. High resolution).

DISCUSSION

The urachus is a fibrous remnant of the allantois, an embryological structure that initially connects the fetal bladder to the umbilicus, facilitating the drainage of urine. In the adult, the urachus typically transforms into a fibrous cord known as the median umbilical ligament. This remnant is located in the space of Retzius, positioned between the transversalis fascia anteriorly and the peritoneum posteriorly. During fetal development, part of the urogenital sinus gives rise to the bladder and the prostatic urethra, while the allantois initially remains open but eventually obliterates as the urachus matures. This anatomical progression is vital to understanding the origin of urachal malignancies.

Urachal cancer, first described by Hue and Jacquin in 1963, is a rare and often aggressive malignancy. It accounts for an estimated 0.01% of all adult cancers and 0.5%-2.0% of all bladder malignancies, representing a significant but infrequent cause of bladder cancer. Urachal adenocarcinoma, in particular, makes up 20%-40% of primary bladder adenocarcinomas, further distinguishing it from more common urothelial bladder carcinomas. The clinical course of urachal carcinoma is typically marked by late presentation, a tendency for early local invasion, and a high propensity for distant metastasis, all contributing to a dismal prognosis. Patients diagnosed with locally advanced or metastatic disease have a mean survival of only 12-24 months, with a 5-year survival rate of approximately 43%.[1] The delayed symptom onset and aggressive behaviour of the disease make early detection and intervention challenging.

Urachal carcinomas primarily arise from the epithelial lining of the urachus. Although the urachal remnant is normally lined with urothelium, these carcinomas often exhibit histological features distinct from conventional bladder cancer. In contrast to most bladder carcinomas, which are predominantly urothelial carcinomas, urachal carcinomas are more often adenocarcinomas. These adenocarcinomas are typically mucin-producing and frequently display focal signet ring cell differentiation. These histological features distinguish urachal adenocarcinomas from typical

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urothelial carcinomas, which usually present as noninvasive papillary tumors that do not invade the muscular wall of the bladder. In contrast, urachal adenocarcinomas tend to infiltrate the muscularis propria and perivesical soft tissues, underscoring their more aggressive nature. Histological subtypes of urachal adenocarcinoma include mucinous, intestinal, and mixed types, with mucinous adenocarcinoma being the most common. Sarcomas, small-cell carcinomas, and transitional cell carcinomas are rare in this context.[2]

One of the major challenges in managing urachal carcinoma is its tendency to remain asymptomatic in the early stages. Symptoms typically develop only when the tumor invades the bladder wall, leading to common clinical signs such as hematuria, irritative voiding symptoms, and mucous-like discharge. Hematuria is present in approximately 90% of patients and is a key indicator of malignancy, increasing the likelihood of cancer by 17-fold, particularly in patients over 55 years of age.[3] Abdominal symptoms such as umbilical pain or discharge may also be reported, particularly in the later stages. These nonspecific symptoms often result in diagnosis at an advanced stage, complicating treatment and limiting the available therapeutic options.[4]

Imaging plays a crucial role in the diagnosis and staging of urachal carcinoma. A combination of ultrasound (US), computed tomography (CT), and magnetic resonance imaging (MRI) is typically employed to assess tumor size, location, and extent of local and distant spread. Ultrasound is frequently the first modality used and can detect the presence of a soft tissue mass with heterogeneity and calcification. Doppler imaging may sometimes reveal internal vascularity, although this is nonspecific. CT scans are particularly useful in visualizing the mixed solid and cystic nature of the tumor, with cystic components often representing mucin. CT also has high sensitivity for detecting calcifications, which are commonly observed in urachal adenocarcinomas. In terms of tumor positioning, 88% of these tumors are found outside the bladder lumen.[5] MRI is valuable for assessing the extent of local invasion, including lymph node involvement, and provides detailed imaging of peritoneal or visceral spread. MRI typically demonstrates high signal intensity on T2-weighted images and post-contrast enhancement, which is attributed to the mucin content and calcifications within the tumor.

Surgical resection remains the cornerstone of treatment for localized urachal cancer. The optimal approach typically involves partial cystectomy along with removal of the urachal ligament and, in many cases, umbilectomy. This combined approach has been shown to result in better outcomes compared to cystectomy alone, as it reduces the risk of recurrence and preserves the patient's quality of life.[6] Studies have consistently demonstrated that this comprehensive surgical approach offers superior survival outcomes. A retrospective case series by Bruins et al,[7] highlighted lymph node metastasis as an independent prognostic factor associated with reduced overall survival (OS). However, there was no significant survival difference between patients who underwent pelvic lymph node dissection (PLND) and those who did not, suggesting that lymph node dissection may not have a significant impact on survival outcomes, at least in the context of this disease.

Recurrence after surgery is common, with reported recurrence rates as high as 40%.[8] This recurrence often manifests within the first two years following surgery, primarily as local recurrence (in the pelvic lymph nodes, peritoneum, or omentum) or distant metastasis, particularly to the lungs and liver.[9] While there is no consensus regarding the use of adjuvant or neoadjuvant chemotherapy, chemotherapy is generally recommended for patients with lymph node involvement, distant metastasis, or positive surgical margins.[10] Radiotherapy is rarely used, with less than 1% of patients receiving it as a monotherapy, and its role remains unclear in the management of urachal carcinoma. The decision to use chemotherapy or radiation therapy should be individualized based on the patient's clinical stage and overall prognosis.

Although follow-up protocols for urachal carcinoma remain somewhat controversial, the high recurrence rate necessitates intensive surveillance. Cross-sectional imaging, particularly CT scans, is commonly employed for routine post-operative surveillance, similar to the follow-up protocol for muscle-invasive bladder cancer. Additionally, cystoscopy is recommended due to the risk of tumor seeding, particularly in patients who have undergone partial cystectomy for noninvasivetumors. Pinthus et al. identified tumor seeding into the lower urinary tract in 8.3% of patients, even after partial cystectomy, suggesting that cystoscopic surveillance is an essential component of post-operative care.[11] Given the significant risk of recurrence and metastatic relapse, an intensive follow-up strategy should be employed, incorporating both imaging and cystoscopy, to ensure early detection of recurrence and to guide subsequent treatment.

CONCLUSION

In conclusion, urachal carcinoma is a rare and aggressive malignancy that poses significant diagnostic and therapeutic challenges. Early detection remains difficult due to its often asymptomatic nature in the early stages. A high level of clinical suspicion, along with comprehensive imaging and histopathological analysis, is essential for diagnosis. Surgical resection remains the mainstay of treatment, with the addition of chemotherapy in cases with advanced disease. Given the high risk of recurrence, close post-operative monitoring, including imaging

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and cystoscopy, is necessary to detect relapses and guide subsequent management.

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