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Mucinous Adenocarcinoma of The Urachus: Case Report and Literature Review

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Abstract

Urachal carcinoma is an uncommon malignant epithelial tumor originating from urachal remnants and often poses a diagnostic challenge, typically being a diagnosis of exclusion. The differential diagnosis of urachal anomalies can be refined by thoroughly assessing factors such as lesion location, morphology, imaging results, patient demographics, and clinical history. In this article, we present a case involving a 54-year-old female with a history of hematuria unaccompanied by other symptoms, ultimately diagnosed with invasive mucinous urachal adenocarcinoma. Our intention with this article is to highlight the pathological features of urachal adenocarcinoma.

Introduction

Adenocarcinomas situated in the bladder region represent a mere 0.5% to 2% of all malignant bladder neoplasms. They can be categorized as bladder primary (non-urachal), originating from remnants of the urachus, or as bladder metastasis [1]. Urachal carcinoma, in particular, is an exceptionally rare and aggressive neoplasm, accounting for less than 1% of bladder cancers and a mere 0.01% of cancers in adults [2,3]. This condition predominantly affects males, typically occurring between the fifth and sixth decades of life [4,5].

This canal closes as the bladder migrates to the pelvis during the fourth month of fetal development, becoming a 5 to 10 cm long fibromuscular vestigial tissue known as the median umbilical ligament. It connects the umbilicus to the bladder, residing within the Retzius (retropubic) space [2].

Autopsy findings reveal that approximately 30% of adults retain a patent lumen within this canal, potentially leading to various anomalies, such as cysts, patent urachus, urachovesical diverticulum, and neoplasms [1,5].

This article aims to present a rare case of mucinous adenocarcinoma originating from the urachus in a female patient. It includes a comprehensive review of the existing literature on this subject.

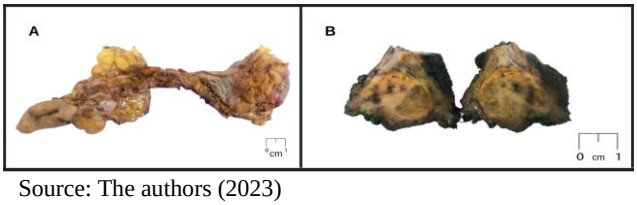
Case Report

A 54-year-old female patient presented with a persistent complaint of hematuria accompanied by clots, which had been ongoing for five months. Upon admission to the hospital, an abdomino-pelvic ultrasound was conducted, revealing an expansive process within the bladder wall involving soft tissue, suspected to be a neoplasm.

To complement the diagnostic investigation, a pelvic magnetic resonance imaging (MRI) was carried out, given the initial findings on the ultrasound, identifying an expansive infiltrative lesion within the bladder. Subsequent radiological staging showed no signs of metastases. During a cystoscopy, the urology team observed an exophytic lesion in the bladder fundus and roof. Transurethral resection biopsies were performed.

Microscopic examination of the biopsy unveiled an invasive adenocarcinoma exhibiting an intestinal pattern. Additionally, a tubulo-villous adenoma and intestinal metaplasia were found in the adjacent mucosa. The clinical data, including the location of the lesion in the bladder fundus/roof and the histopathological characteristics identified, raised the possibility of a urachal origin. This suspicion emerged after excluding the likelihood of the neoplasm originating from the colon, as endoscopic/colonoscopic examinations revealed no gastrointestinal tract lesions.

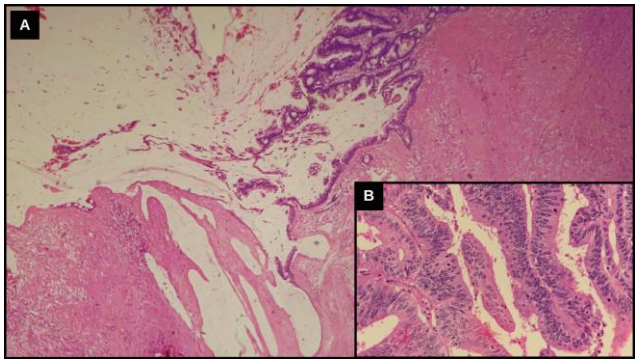
Subsequently, it was decided to resect the urachal neoplasm and perform a partial cystectomy. The surgical specimen comprised an elliptical section of skin at one end, bearing an umbilical scar, and an area of cystectomy at the opposite end with a granular appearance and a focus of ulceration (Figure 1A). Sections from the specimen displayed a heterogeneous lesion centered on the bladder wall (Figure 1B).



Source: The authors (2023)

Figure 1: A. Surgical specimen from urachal tumor resection measuring 15.3 × 4.6 cm at its longest axes, with one end containing the umbilical scar and the contralateral end involving the bladder region. B. A heterogeneous, well-defined lesion with a variegated coloration, ranging from yellow-orange to whitish-brown, measuring 3.5 × 3.0 cm, primarily located on the bladder wall.

Further microscopic analysis of the surgical specimen confirmed the diagnosis of invasive mucinous adenocarcinoma of urachal origin, characterized by a moderately differentiated intestinal pattern (Figure 2A). This lesion extended into the muscularis propria layer (detrusor muscle) and infiltrated the bladder mucosa. An adjacent precursor lesion of the villous adenoma type originating from the urachus was also identified (Figure 2B). No angiolymphatic or perineural invasion was detected, and the surgical margins were free from neoplastic involvement. Pathological staging, as per the Sheldon system, classified the tumor as stage IIIA.



Source: The authors (2023).

Figure 2: A - Invasive mucinous lesion in the adjacent tissue (Optical Microscopy, Hematoxylin-Eosin, 100x). B - Precursor lesion of villous adenoma with high-grade dysplasia, noticeable nuclear atypia (Optical Microscopy, Hematoxylin-Eosin, 400x).

Discussion

Urachal carcinoma is a rare malignant epithelial tumor originating from urachal remnants and often poses a diagnostic challenge, typically being a diagnosis of exclusion. It predominantly manifests on the bladder dome, with rarer occurrences along the midline of the anterior wall or above the umbilicus [6]. Initial symptoms are non-specific, with hematuria being the most common, followed by abdominal pain, dysuria, and mucosuria [5].

The incidence of urachal cancer is exceptionally low, estimated at less than 1 case per 1 million person-years. It typically emerges during the fifth or sixth decade of life, although cases span a wide age range. The male-to-female ratio stands at approximately 1.2-1.4:1 [7]. The majority of cases fall under the histological subtype of non-cystic adenocarcinomas, with various subtypes including enteric, mucinous, signet ring cell, and mixed [8]. Uncommon non-glandular variants encompass urothelial, squamous, and neuroendocrine carcinomas [9].

The primary etiological factor behind these neoplasms is thought to be the potential glandular metaplasia of the transitional epithelium lining the urachal remnant, which may predispose it to adenocarcinoma [10]. Macroscopically, solid cases typically present as firm, whitish or greyish infiltrative tumors, while cystic variants also exist [11]. Diagnostic imaging methods include ultrasound, revealing an irregular, heterogeneous mass in the midline supravesical area, and computed tomography and magnetic resonance imaging, which can assess lesion extension, lymph node involvement, and distant metastasis. Calcifications in the bladder, particularly in the midline, raise strong suspicions of urachal origin. Cystoscopy is useful for tumor localization and facilitates biopsy [12].

Diagnostic criteria for urachal adenocarcinoma include tumor location in the anterior wall or dome of the bladder, epicenter of the carcinoma within the bladder wall, the absence of cystitis with a generalized cystic or glandular pattern in the dome or anterior wall of the bladder, absence of primary adenocarcinoma elsewhere, and origination from a urachal remnant [4,5].

Histopathologically, non-cystic enteric adenocarcinoma closely resembles colorectal adenocarcinoma, displaying glandular and cribriform architecture, nuclear pseudostratification, and necrosis. The mucinous subtype features extracellular mucin lakes, sometimes with colloid-type mucin dissecting the stroma and the presence of signet ring cells floating within the mucin reservoirs. The signet ring cell subtype is reserved for tumors with extensive permeative stromal growth. Non-glandular carcinomas share morphological similarities with their counterparts in the bladder [13].

Solid urachal adenocarcinomas typically exhibit positivity for CDX2 and CK20, along with nuclear β-catenin staining, while GATA3 is not expressed [12]. The Sheldon system, established in 1984 and categorized into four stages (Table 1), is widely used for staging urachal adenocarcinomas [12].

Sheldon System for Staging Urachal Adenocarcinomas		
I	Tumor confined to the mucosa	
II	Invasive tumor confined to the urachus	
III	Invasion through the urachus	A - Extension into the bladder
		B - Extension into the abdominal wall
		C - Extension into the peritoneum
		D - Extension to other organs (except the bladder)
IV	Metastases	A - Lymph node metastases
		B - Distant metastases

Table 1: Sheldon system for staging urachal adenocarcinomas.

Metastases are relatively common, primarily affecting the bones, lungs, liver, non-regional lymph nodes, and peritoneum. Multiple-site metastatic disease and local recurrence are prevalent [14]. The average 5-year survival rate is approximately 50%, which is higher compared to adenocarcinomas of equivalent stage or urothelial carcinomas of the urinary bladder [15].

Treatment for non-metastatic urachal cancer typically involves partial or radical cystectomy, with partial cystectomy preserving patients' quality of life and yielding similar outcomes. Chemotherapy is an option in cases of metastatic disease or local recurrence, with treatment regimens based on methotrexate, vinblastine, and doxorubicin or 5-fluorouracil5.

Conclusion

Urachal adenocarcinoma is an aggressive neoplasm that arises from embryological remnants, considered a rare condition with a varied clinical presentation, which may include symptoms such as hematuria, suprapubic pain, and thickening of the bladder dome on imaging studies. The primary approach to treatment involves surgical intervention. Excluding the primary site within the gastrointestinal tract is crucial, given the location and microscopic appearance of these neoplasms.

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Conflict of Interest

None of the authors have any conflicts of interest to disclose.

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