

Primary Urachal Adenocarcinoma with signet ring cell carcinoma: a rare case report

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Abstract: The primary urachal carcinoma is a very rare malignant tumor. It generally presents as a high grade, high stage tumor and in most of the cases it has regional or distant metastasis at the time of presentation. It is known to have a uniformly poor prognosis. In this paper, we present here a very interesting and rare case report of primary urachal adenocarcinoma with signet ring cell carcinoma in a 58-year-old male who presented the lower abdomen has a mass and felt discomfort. In this case, urachal carcinoma were successful treated by surgery using partial bladder cystectomy with excision of the urachal mass and umbilicus, and the patient also underwent systematic chemotherapy. Followed up for 10 months, the patient has not experienced recurrence or other metastasis. Organ preserving partial cystectomy provides higher quality of the life and chemotherapy is the primary method used to treat the disease.

Keywords: Urachal carcinoma; Urachus; Therapy

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1. Introduction

The urachus is a tubular structure connecting urinary bladder and the umbilicus. As the main excretory organ in fetus, the urachus moves the nitrogenous waste from the bladder. During the fourth and fifth month in embryonic life, the urachus gradually degenerates into a rudimentary fibromuscular closed canal, which is known in adults as the median umbilical ligament and stretches between the dome of urinary bladder and the umbilicus. However, urachal remnant in the form of a tubular or cystic muscular structure can persist. It is most commonly found attaching the umbilicus to the bladder at the dome. Failure of complete urachal lumen closure may lead to various anomalies including infection, cystic formation and malignant transformation[1,2]. Autopsy studies suggested that in one-third of adults the urachus canal partly persists[3]. Urachal carcinoma (URC) is a rare malignant disease that is located in the residual urinary tissue, accounted for 0.01% of adult tumors and about 20-39% of primary bladder adenocarcinoma. Urachal tumors tend to be associated with a poor prognosis, with 5-year survival rates ranging from 9.0 to 43% [4]. In this paper, a patient who treated URC in December 2016 is introduced the diagnosis and treatment are discussed.

2. Materials and Methods

2.1. Case report

A 58-year-old male, no smoking history, was found to have a lump approximate size 5.0×3.0cm at the lower abdomen five years ago and the lump gradually increasing one year and felt discomfort. There was no symptoms of hematuria and irritation of the bladder, also no history of weight loss. The patient had a history of hypertension about 24 years, regular to take medication the blood pressure control smoothly.

On physical examination, a defined lump was

palpable in the umbilical and hyponastic region, globular in shape, of approximate size 12×10cm, had smooth surface and regular margins all around. The rest of the physical examination was absolutely normal. Ultrasonography showed a solid and cystic heterogenous mass of size about 12.4x10.1x9.2cm above urinary bladder, Internalultrasound image is uneven and had a sheet of high ultrasound image inside, it close to the left wall of the bladder, with well bladder filling, smooth mucosa and no obvious significant mass. Urine microscopy and urine cytology were absolutely normal. Computed Tomography (CT) of the abdomen showed a hypodense soft tissue density lesion in supravescical region from the dome of urinary bladder and the lesion extended superiorly up to just below the umbilicus, so the border between the lower margin and the bladder is not clear (Figure 1). There was no evidence of distant metastasis or intra-abdominal lymphadenopathy and the other organs appeared normal. Provisional diagnosis made was urachal remnant malignancy or the carcinoma urinary bladder. The patient underwent cystoscopic examination, which showed that at the anterior and dome of the bladder there is a round swollen squeezing the urinary bladder, the bladder mucosa was smooth. Table 1 summarizes the clinicopathological features of this case.

2.2 Therapy

The patient underwent surgical exploration and urachal mass extending up to the dome of urinary bladder and superiorly just below umbilicus was found. Partial bladder cystectomy with excision of the urachal mass and umbilicus was done and confirming the absence of leakage from the urinary bladder. After the operation, the patient carried out systematic chemotherapy.



Figure 1. Computed Tomography of abdomen showing urachal mass (arrow) attaching the bladder dome and squeezed the bladder.

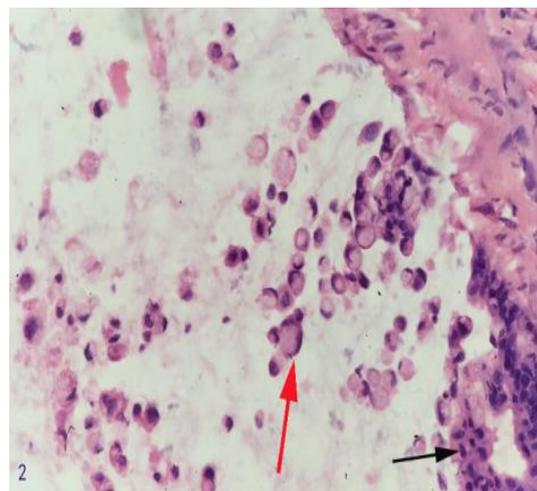


Figure 2. Histopathological findings of the biopsy showing the tumour cells containing intracellular vacuole displacing the hyperchromatic nucleus to one side suggestive of signet ring cell carcinoma (red arrow), urachal mucinous adenocarcinoma (black arrow).

Table 1 Clinicopathological factors of this patient

Factors Patient	
Age	58
Gender	Male.
Symptoms	Abdomen discomfort.
Surgery	Partial bladder cystectomy with excision of the urachal mass and umbilicus.
Location	Dome of the bladder.
Size	12.4x10.1x9.2cm.
Histology	Adenocarcinoma and signet ring cell carcinoma.
CC/CG	None.
Urachal remnants	Yes.
Myao stage at diagnosis	I.

CC cystitis cystica, CG cystitis glandularis.

3. Results

Pathological results showed: urachal mucinous adenocarcinoma, include a part of the signet ring cell carcinoma, invasion of the full urachus layer and the inherent muscular layer of the bladder is infiltrated (Figure 2). The patient has been regular followed up for 10 months and doing is well.

4. Discussion

URC is an extremely rare but highly malignant entity representing 0.35%-0.70% of all bladder cancers [5, 6]. Primary urachal adenocarcinoma is a rare tumor, accounting for only 0.17–0.34 % of all bladder tumors[7, 8]. Hue and Jacquin in 1863 first described the urachal cancer. To date, no consensus has been reached regarding the diagnostic criteria of URC. The most commonly used criteria proposed by Sheldon et al [7] and Mostofi et al[9] and revised by Gopalan et al[5], which includes the following characteristics: (1) tumor is located in the dome/anterior wall of the

bladder, (2) epicenter in the bladder wall, (3) absence of cystitis cystica and cystitis glandularis, and (4) lack of known primary adenocarcinoma elsewhere. The most frequent symptom of URC is macroscopic or microscopic hematuria followed by abdominal pain and dysuria. Other less frequent clinical presentations included pollakisuria, pyuria, urinary tract infection, umbilical discharge (e.g., blood, urine, and mucus), vaginal discharge, and nonspecific symptoms (nausea, vomit, diarrhea, weight loss, or fever)[10]. Some serum markers have proven to be helpful in the diagnosis and monitoring of URC, carcinoembryonic antigen and carbohydrate antigen19-9 as well as cancer antigen 125 serum levels have been detected to be increased in URC adenocarcinoma[11,12]. In this case, the patient presents with a mass of the abdomen, no hematuria and abdominal pain, the serum markers appeared normal. When a patient presents with non-specific symptoms like abdomen mass in our case, then a high index of suspicion is required because these symptoms are very common with benign

conditions. The most common histological type of URC is adenocarcinoma. Other rare patterns include the signet ring cell type, clear cell type, hepatoid type and mixed patterns[13]. It has been postulated that the oncogenesis of urachal adenocarcinoma involves a metaplastic process, as the urachal urothelium often exhibits glandular metaplasia[5]. Several types of stage classifications are suggested, but the most often used are the Mayo staging systems, which includes the following[7]: I. Tumor confined to urachus and/or bladder, II. Tumor extending beyond the muscular layer of urachus and/or the bladder, III. Tumor infiltrating the regional lymph node, IV. Tumor infiltrating non-regional lymph nodes or other distant sites. URC generally present as high grade, high stage and have a uniformly poor prognosis and in the majority of cases there are regional or distant metastasis at the time of presentation[14]. By lymphatic dissemination URC usually metastasizes into the pelvic lymph nodes and by hematogenous dissemination into distant organs, especially lung, bone, or peritoneum[15]. The recommended treatment for nonmetastatic URC is surgery. Some scholars believe that both partial and radical cystectomy can be considered as they provide similar oncological results[16]. However, organ preserving partial cystectomy provides higher quality of life and should therefore be preferred. URC is not sensitive to radiotherapy, chemotherapy is the only treatment option to potentially prolong survival. Meta-analysis shows that the most effective treatment may be combination of 5-FU with cisplatin, which performs significantly better than cisplatin-based therapies[10].

In summary, URC is an extremely rare cancer and often presents as a high grade, high stage. But the localized URC organ preserving partial cystectomy provides a long-term disease-free survival and the combination of 5-FU with cisplatin provides the most favorable response. On the other hand, the postoperative regular follow-up is necessary.

References

- [1] Irwin PP, Weston PMT, Sheridan W, et al., Transitional cell carcinoma arising in a urachal cyst[J]. *Bju International*. 2010, 67(1):103-104.
- [2] Negro F, Crucitti A, Urachal adenocarcinoma. A case report[J]. *Minerva Chirurgica*. 1994, 49(9):885-888.
- [3] Wilcox DT, Godbole PP, Koyle MA. *Pediatric Urology: Surgical Complications and Management* 2009.
- [4] Henly D, Farrow G, Zincke H, et al., Urachal cancer: the role of conservative surgery[J]. 1993.
- [5] Gopalan A, Sharp DS, Fine SW, et al., Urachal carcinoma: a clinicopathologic analysis of 24 cases with outcome correlation[J]. *American Journal of Surgical Pathology*. 2009, 33(5):659.
- [6] Johnson DE, Hodge GB, FW A-K, et al., Urachal carcinoma[J]. *Urology*. 1985, 26(3):218-221.
- [7] Sheldon CA, Clayman RV, Gonzalez R, et al., Malignant urachal lesions[J]. *Journal of Urology*. 1984, 131(1):1.
- [8] Fanning DM, Sabah M, Conlon PJ, et al., An unusual case of cancer of the urachal remnant following repair of bladder exstrophy[J]. *Irish Journal of Medical Science*. 2011, 180(4):913-915.
- [9] Mostofi FK, Thomson RV, Dean AL, Mucous adenocarcinoma of the urinary bladder[J]. *Cancer*. 1955, 8(4):741-758.
- [10] Szarvas T, Módos O, Niedworok C, et al., Clinical, prognostic, and therapeutic aspects of urachal carcinoma-A comprehensive review with meta-analysis of 1,010 cases[J]. *Urologic Oncology*. 2016, 34(9):388-398.
- [11] Hayashi T, Yuasa T, Uehara S, et al., Clinical outcome of urachal cancer in Japanese patients[J]. *International Journal of Clinical Oncology*. 2016, 21(1):133-138.
- [12] Siefker-Radtke AO, Gee J, Shen Y, et al., Multimodality management of urachal carcinoma: the M. D. Anderson Cancer Center experience[J]. *Journal of Urology*. 2003, 169(4):1295-1298.
- [13] Williamson SR, Lopez-Beltran A, Montironi R, et al., Glandular lesions of the urinary bladder: clinical significance and differential diagnosis[J]. *Histopathology*. 2011, 58(6):811-834.
- [14] Wood DP, *Urothelial Tumors of the Bladder*[J]. 2012.
- [15] Chen ZF, Wang F, Qin ZK, et al., Clinical Analysis of 14 Cases of Urachal Carcinoma[J]. *Chinese Journal of Cancer*. 2008, 27(9):966-969.
- [16] Bruins HM, Visser O, Ploeg M, et al., The clinical epidemiology of urachal carcinoma: results of a large, population based study[J]. *J Urol*. 2012, 188(4):1102-1107.