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Case Report

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Urachal Carcinoma: A Case Report of Mixed Adenocarcinoma with Squamous Cell Carcinoma and Literature Review

LI Hongbo[1], CAI Fei[2] , ZHANG Kai[1], QIAO Xubai[2], GU Yanfei[3], Francisco J. Martinez Portillo[1], ZHU Gang[1]

[1] Department of Urology, Beijing United Family Hospital, Beijing 100015, China.

[2] Department of Pathology, Beijing United Family Hospital, Beijing 100015, China.

[3] Department of Oncology, Beijing United Family Hospital, Beijing 100015, China

*Corresponding author: Zhu Gang, Department of Urology, Beijing United Family Hospital, Beijing 100015, China

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Abstract

Primary urachal carcinomas are very rare urological malignancies with poor prognosis. While adenocarcinoma is common among urachal carcinomas, whereas squamous cell carcinoma (SCC) is very rare. We reported a case of a mixed adenocarcinoma with squamous cell carcinoma arising in the urachus with literature review.

Keywords: Adenocarcinoma; Mixed urachal carcinoma; Squamous cell carcinoma; Urachal carcinoma

Introduction

Urachal carcinomas is a rare aggressive tumor which arises from the urachal remnants. The incidence of urachal carcinoma is one in 5 million or 0.01% of all cancers in adults [1]. It is most commonly found at the bladder dome but can present in the anterior or posterior wall and extend to the umbilicus. The most common pathology type of urachal carcinoma is adenocarcinoma, which believed to evolve from intestinal metaplasia of the epithelium of the urachal remnant [2]. Squamous celland anaplastic carcinomas also arise from the urachus [3]. The non-glandular types including urothelial, squamous cells, neuroendocrine, and mixed type which are very rare.

We report a urechal carcinoma case with mixed adenocarcinoma with squamous cell carcinoma.

Case Report

A 35-year-old man, who was previously well, presented to Emergency Room for painful urination and fresh blood clot in his urine in Sept. 2017. He was with urinary frequency and urgency about 6 months ago and treated as Urine Tract Infection (UTI). He also had some on and off lower abdominal pain radiating to the bilateral inguinal area. The pain score was 5-6/10. Cystoscopy in local hospital found a lesion in the bladder dome. Biopsy showed squamous cell carcinoma. His biopsy specimen slides had been reviewed by our pathologists. The histological diagnosis confirmed squamous cell carcinoma, moderately differentiated. The immunohistochemical study showed GATA-3 (Negative), P40 (Positive), CK20 (Scattered positive).

He was a smoker for over 10 years, with 10 cigarettes per day. No cancer family history. The physical exam found a soft abdomen without tenderness, no enlarged lymph node in the inguinal area. CT scan with contrast showed a 3.5×3.5×4.0 cm mass in the midline at the dome of the bladder with calcification, involving the bladder mucosa at the dome as showing in Figure 1. 3D image reconstruction of the CT scan was performed and showed the size, location, and blood supply to the tumor as showing in Figure 2.

Figure 1: CT scan found a mass at the dome of bladder with calcification.

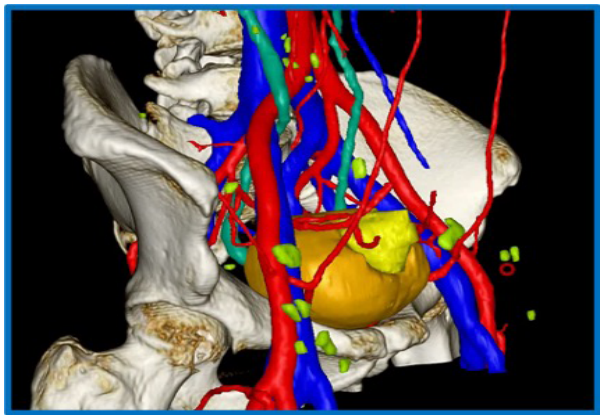

Figure 2: 3D image reconstruction based on CT scan.

Based on the clinical results, the diagnosis of urachal carcinoma was established. Robotic-assisted laparoscopic partial cystectomy, urachus excision and bilateral pelvis lymphadenectomy were performed on 2017.11.07. The umbilicus was excised with a connection to the urachus. The urachus excision was conducted to approach the bladder dome. A partial cystectomy was performed to achieve the complete excision of the urachal tumor. Flexible cystoscopy was performed simultaneously in the procedure in guiding the resection border as showing in Figure 3.

Figure 3: Flexible cystoscopy view of the urachal tumor.

All the samples at 3, 6, 9, 12 clockwise were negative in the frozen section histology examination. The bladder defect was closed with 2-0 V-Loc sutures in two layers fashion. Bilateral pelvic lymphadenectomy was conducted. There were enlarged lymph nodes noticed bilaterally. The Estimated Blood Loss (EBL) was 50 mls. The operation time was 90 minutes. The patient recovered uneventfully and was discharged at day 7 post-operatively Figure 4.

Figure 4: The umbilicus was excised with a connection to the urachus and tumor.

Histopathological findings

Big slide technique was applied in this case (Figure 5). Histopathological examination showed a mixed type of carcinoma (Figure 6). Most of the tumor is a moderately differentiated adenocarcinoma (Figure 7). The grossly white hard area is a well-differentiated squamous cell carcinoma (Figure 8) accounting for about 5%. The tumor was distinct from the lining of the bladder, which was histologically normal. On immnohistochemical study, the adenocarcinoma component of the tumor was positive for CD7, CD20, and CDX2. β-catenin membranes positive while a nuclear negative (Figure 9) showed a lack of nuclear localization in contrast to metastatic colonic adenocarcinoma. The squamous cell carcinoma component of the tumor was positive for CK5/6 and P40 (Figure 10).

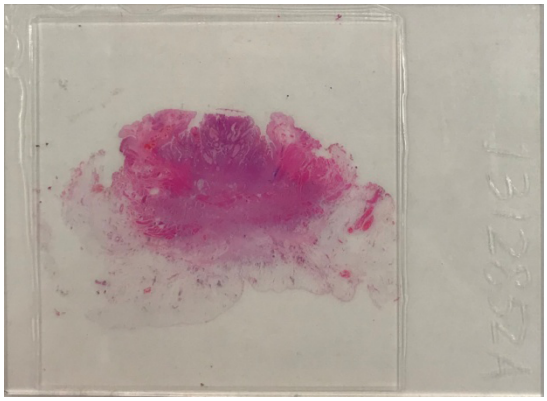

Figure 5: Big slide of specimen cell type.

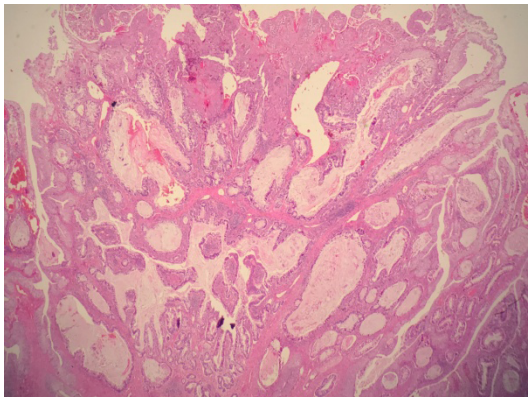

Figure 6: Histopathology exmination showed mixed.

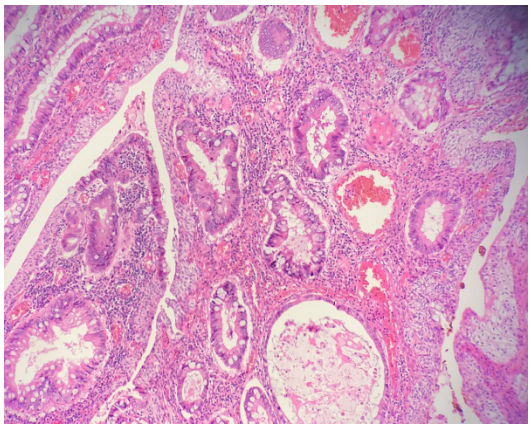

Figure 7: Most of the tumor is a moderately differentiated adenocarcinoma.

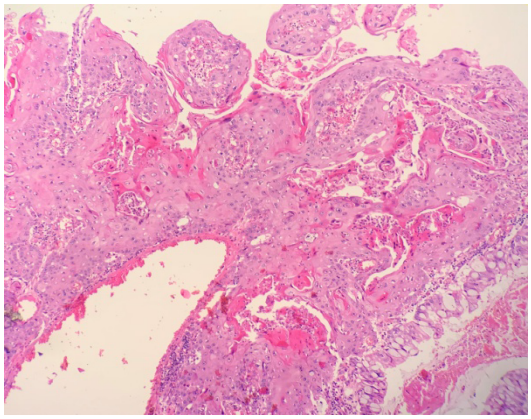

Figure 8: The grossly white hard area is a well-differentiated squamous cell carcinoma.

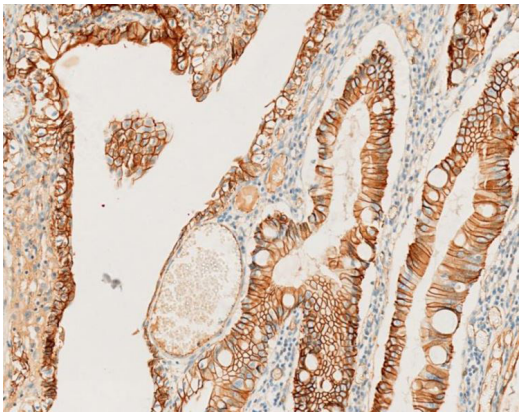

Figure 9: Immnohistochemical study, the adenocarcinoma component of the tumor was positive for CD7, CD20, and CDX2. β-catenin membranes positive while a nuclear negative.

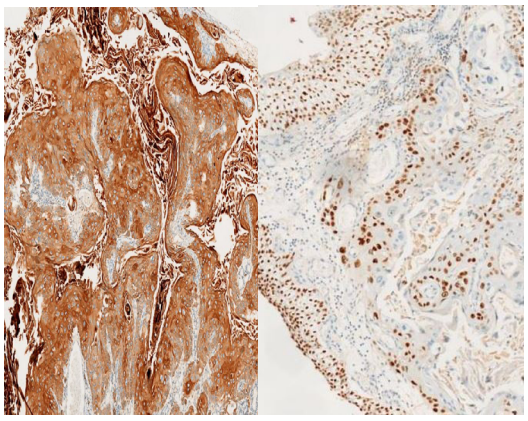

Figure 10: The Squamous cell carcinoma component of the tumor was positive for CK5/6 and P40.

Presence of perineural invasion was found. Surgical margins were uninvolved by carcinoma. Bilateral para-umbilicus artery lymph nodes showed no malignancy.

MDT for treatment plan

A MDT attended with urologists, oncologists, and radiotherapiest was conducted after the surgery in our insititue. As there was no local lymph node invasion and metastasis, histopathology showed complete excision of the primary tumor, we did not recommend adjuvant chemotheapy or radiotherapy after the MDT discussion. He was followed up every 3 months with abdomen and pelvis MRI, Chest X-ray every 6 months in the first year. Then the follow up plan will be adjusted according to patient's condition.

Follow up results

Patient was followed up regularly. He was found a 2.8X2.7X2.9cm mass connected with the anterior wall of the mid-upper rectum and behind the seminal vesicle, more likely metastasis by MRI on 2018.11.08. He then accepted robotic tumor excision and rectectomy done by general surgeon and then accepted chemotherapy and radiotherapy by oncologist. Up to the date of this paper submission, patient was recurrence and metastasis free.

Discussion

The urachus is a thick tube-like structure that is formed in the embryo as the allantois involutes. It extends from the umbilicus to the bladder dome. After birth, it becomes a fibrous cord called the median umbilical ligament. If remnants of the allantois remain within the ligament, they develop into cysts as well as epithelial neoplasms. A urachal remnant may persist in approximately 32% of adults [4], consisting of a tubular or cystic structure lined by epithelium, surrounded by connective tissue and musculature. The epithelial lining of the urachus is urothelium, similar to that of the urinary bladder and the ureter, but it frequently undergoes metaplastic change, mostly glandular. An epithelium-lined lumen usually persists throughout life and uncommonly gives rise to aggressive urachal carcinoma [3].

The pathologic criteria for the diagnosis of urachal carcinoma are varied. Criteria for diagnosis of urachal adenocarcinoma accepted by the newest WHO as follows: 1. Location of the tumor in the bladder dome and/or anterior wall; 2. The epicenter of carcinoma in the bladder wall; 3. Absence of widespread cystitis cystica and/or cystitis glandularis beyond the dome or anterior wall; 4. Absence of a known primary elsewhere [5].

Several staging systems have been used for urachal carcinomas. No AJCC TNM staging system for urachal carcinomas as urachal tumors do not arise from the bladder surface urothelium [6]. The most widely used staging system is the Sheldon system [7].

The Sheldon staging system for Urachal carcinoma. Stage I: Carcinoma confined to the Urachal mucosa; Stage II: Carcinoma invasion confined to the urachus; Stage III: Local carcinoma extension, IIIA Extension into the bladder. IIIB Extension into the abdominal wall. IIIC Extension into the peritoneum. IIID Extension into other viscera; Stage IV: Metastasis. IVA Metastasis to lymph nodes. IVB Metastasis to distant sites. In this case, it was Sheldon IIIA. The mixed type of urachal carcinoma is rare. There were only five cases reported with our knowledge. Surgery is the primary treatment for urachal carcinomas. There is no standard neoadjuvant or adjuvant chemotherapy. Urachal carcinomas occur mostly in the bladder dome and are generally treated by partial cystectomy with en bloc resection of the medial umbilical ligament and umbilicus [8]. Detailed pathologic studies with clinical outcome correlation are few. The value of the 3D image reconstruction of CT scans in this case helped surgeon understand the blood supply and tumor boundary well before surgery and assist the surgical procedure plan. This can be beneficial for the decision making in the range of tumor margin excision, in lymphadenectomy as well as the hemostasis in the operation.

References

1. Scabini S, Rimini E, Romairone E, et al. (2009) Urachal tumour: case report of a poorly understood carcinoma. World J Surg Oncol 7: 82.
2. Wang D, Sule N (2019) Mucinous Cystadenoma of the Urachus and Review of Current Classification of Urachal Mucinous Cystic Neoplasms. Arch Pathol Lab Med 143: 258-263.
3. Gopalan A, Sharp DS, Fine SW, et al. (2009) Urachal carcinoma: a clinicopathologic analysis of 24 cases with outcome correlation. Am J Surg Pathol 33: 659-668.
4. Paner GP, Lopez-Beltran A, Sirohi D, Amin MB (2016) Updates in the Pathologic Diagnosis and Classification of Epithelial Neoplasms of Urachal Origin. Adv Anat Pathol 23: 71-83.
5. Amin M B ESB, Greene F L, et al. (2016) AJCC Cancer StagingManual, 8th edition. New York: Springer 2016: 739-767.
6. Sheldon CA, Clayman RV, Gonzalez R, Williams RD, Fraley EE (1984) Malignant urachal lesions. J Urol 131: 1-8.
7. Elser C, Sweet J, Cheran SK, Haider MA, Jewett M, et al. (2012) A case of metastatic urachal adenocarcinoma treated with several different chemotherapeutic regimens. Can Urol Assoc J 6: E27-E31.
8. Williams CR, Chavda K (2015) En Bloc Robot-assisted Laparoscopic Partial Cystectomy, Urachal Resection, and Pelvic Lymphadenectomy for Urachal Adenocarcinoma. Rev Urol 17: 46-49.