Case Report

A case of mucosal-associated lymphoid tissue lymphoma of the urachus

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Abbreviations & Acronyms
MALT = mucosa-associated lymphoid tissue
PSA = prostate-specific antigen
RT = radiotherapy

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Introduction: Urachus carcinoma is a rare malignancy with an aggressive potential and a poor prognosis, and evidence is limited for its diagnosis and treatment.
Case presentation: A 75-year-old man underwent fluorodeoxyglucose positron emission tomography/computed tomography for staging prostate cancer, and a mass (standardized uptake value max 9.5) was observed on the outside of the urinary bladder dome. T2-weighted magnetic resonance imaging showed the urachus and a low-intensity tumor, which suggested a malignant tumor. We suspected urachal carcinoma and performed total resection of the urachus and partial cystectomy. Pathological examination revealed mucosa-associated lymphoid tissue lymphoma with cells positive for CD20 and negative for CD3, CD5, and cyclin D1. After the surgery, no recurrence has been observed for more than 2 years.
Conclusion: We encountered an extremely rare case of mucosa-associated lymphoid tissue lymphoma of the urachus. Surgical resection of the tumor provided an accurate diagnosis and good disease control.

Key words: bladder cancer, malignant lymphoma, MALT lymphoma, urachal cancer, urachal remnant.

Keynote message

Urachal tumors are rare, which makes their preoperative diagnosis challenging. Surgical resection for localized tumors can be an effective way to accurately diagnose and treat localized tumors with curative intent.

Introduction

The urachus is an embryological remnant that connects the umbilicus and anterior wall of the bladder. During the fourth or fifth months of embryonic development, the urachus is gradually occluded as a fibrous cord.[1] However, up to 30% of the human urachus is not completely occluded before birth and may persist into adulthood. Urachal abnormalities are rare, and common complications include malignant tumors or infections. MALT lymphoma is a subtype of extranodal lymphoma that accounts for 7%–8% of B-cell lymphomas and is associated with chronic inflammation. The correlation between *Helicobacter pylori* and gastric MALT lymphoma is well-known.[2] However, MALT lymphoma rarely affects the genitourinary organ.[3] Here, we report an extremely rare case of MALT lymphoma arising in the urachus.

Case presentation

A 75-year-old man was referred to our department with an elevated serum PSA level (8.14 ng/mL). He had no medical history related to the urachus. Transrectal prostate needle biopsy revealed prostate cancer, with a pathological type of adenocarcinoma and a Gleason score of 8. Fluorine-18 fluorodeoxyglucose positron emission tomography/computed

tomography was used for staging, which revealed a 30-mm mass protruding from the dome of the bladder to the central lower abdominal wall (standardized uptake value max 9.5; Fig. 1). T2-weighted magnetic resonance imaging revealed a low-intensity mass in the dome of the bladder along the urachus (Fig. 2), which was suspected to be a malignant tumor. The periphery was well-defined and unlikely to be invasive to the surrounding tissues. Additionally, there were no abnormalities that were suspected to be metastatic lesions of prostate or urachal cancer. Cystoscopy revealed elevated mucosa in the dome of the bladder (Fig. 3). We diagnosed the patient with localized urachal cancer and decided to perform total resection of the urachus and partial cystectomy. Simultaneously, we diagnosed localized prostate cancer and initiated hormonal therapy. The operating time was 76 min, and minimal bleeding occurred during surgery. The tumor did not adhere to the surrounding tissues and showed good mobility. However, en bloc resection of the bladder wall was performed because the possibility of a malignant tumor could not be excluded during surgery. Microscopic findings showed fibrous tissue that was considered as the urachus and diffuse proliferation of lymphocytes (Fig. 4a). The pathological examination, which used immunohistochemical staining, showed that the cells were positive for CD20 but negative for CD3, CD5, and cyclin D1 (Fig. 4b). The lesion was found to exist in the extranodal marginal zone, without any high-grade features. B cells showed diffuse proliferation without any follicle formation, ruling out the possibility of mantle lymphoma or follicular lymphoma. Based on these findings, the diagnosis of MALT lymphoma was made. The patient has not experienced any postoperative complications or evidence of recurrence for more than 2 years. The serum PSA level was maintained below the detectable level under hormonal therapy.

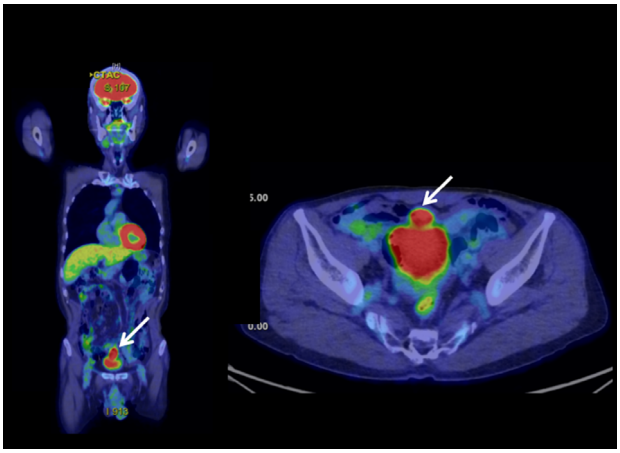

Fig. 1 As the arrows point out, Fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography shows a mass with a hot spot (standardized uptake value max 9.5) protruding from the dome of the bladder to the central lower abdominal wall.

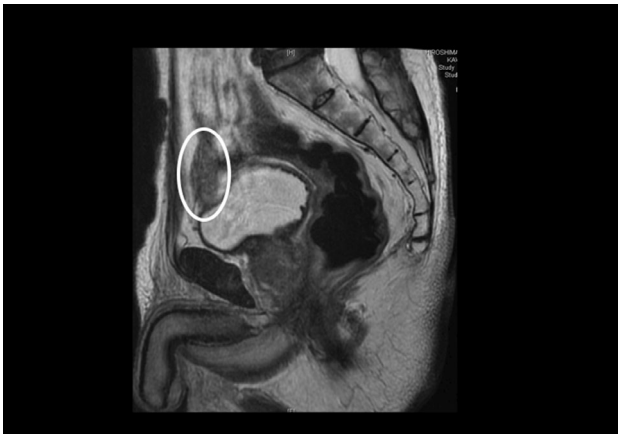

Fig. 2 As the circle points out, T2-weighted magnetic resonance imaging shows a mass with a low-intensity area from the dome of the bladder along the urachus.

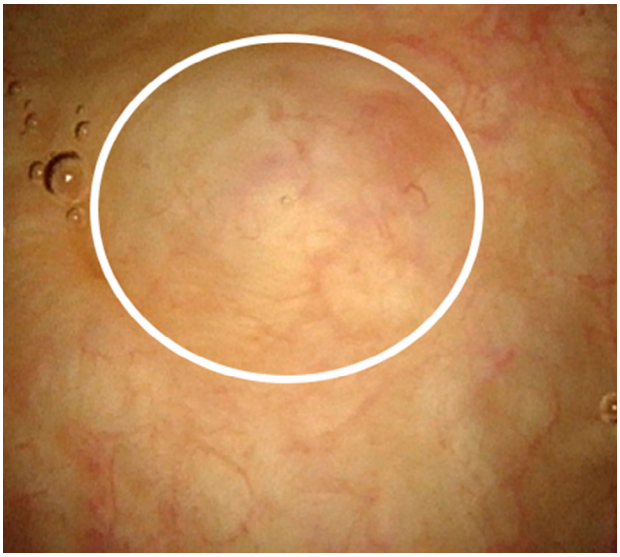

Fig. 3 As the circle points out, cystoscopy shows elevated mucosa on the posterior wall of the bladder.

Discussion

Urachal carcinomas are rare malignancies, accounting for approximately 1.0% of urinary bladder cancer.[4] Therefore, the evidence regarding diagnosis and treatment is sparse, and clinical features, typical imaging findings, and effective tumor markers remain unknown.[5] Diagnostic criteria include the location of the tumor in the dome or anterior wall of the bladder, growth of the tumor in the bladder wall, absence of atypical cystitis or glandularis beyond the dome/anterior wall, absence of urothelial carcinoma in the bladder, and absence of another origin of adenocarcinoma.[6] Although the patient was diagnosed with prostate adenocarcinoma, we excluded the possibility of metastasis of prostate cancer because his initial PSA level was low, and the urachus is an uncommon site for solitary metastasis of prostate cancer.

Urachal carcinomas typically have aggressive potential[5]; therefore, rapid diagnosis and treatment are necessary.

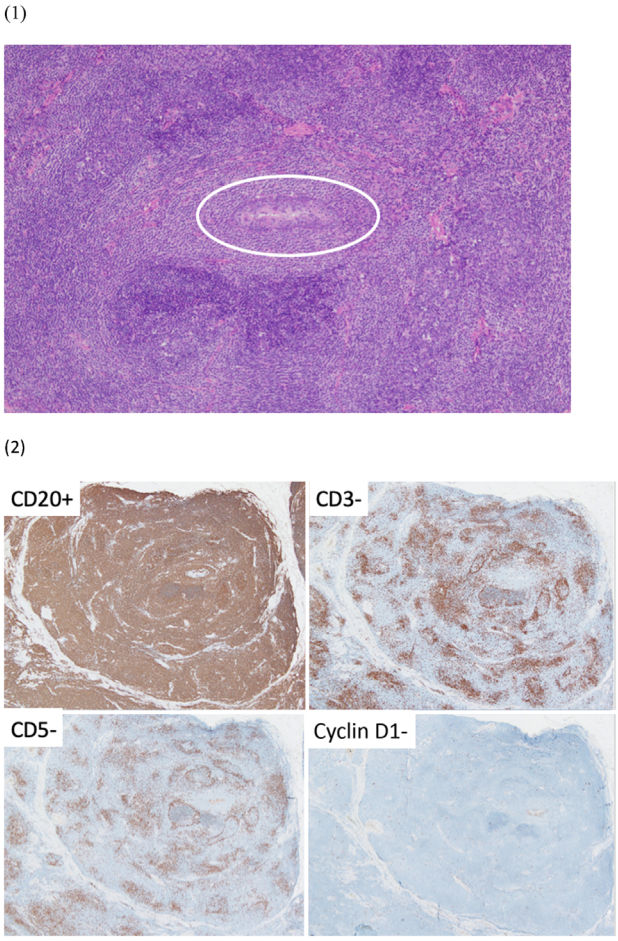

Fig. 4 (1) On microscopic findings, the circle points out a lumen structure considered to be the urachus (magnification ×100). (2) Pathological findings of immunohistochemistry staining show cells that are positive for CD20 and negative for CD3, CD5, and cyclin D1 (magnification ×20).

However, detecting tumors localized in the urachus can be challenging, and patients may not experience objective symptoms or abnormalities in urine tests or cytology. Additionally, transurethral tissue sampling may result in bladder wall perforation when tumors do not grow inside the bladder cavity. Percutaneous needle biopsy carries the risk of puncturing adjacent organs, such as the small intestine. Furthermore, there are no specific pathological criteria for diagnosing urachal carcinomas, although adenocarcinoma is the typical pathology.[7] Therefore, under clinical suspicion of localized urachal carcinoma, tumor removal is acceptable, even without pathological confirmation. The previous report showed that two rare tumors arose in the urachus, namely a desmoid and a solitary fibrous tumor.[8] The preoperative diagnosis for both tumors was urachal carcinoma, and frozen sections of the intraoperative biopsy revealed no malignancy. The report concluded that an accurate preoperative diagnosis was difficult.[8]

To the best of our knowledge, this is the first reported case of urachal MALT lymphoma. MALT lymphoma is a subtype of extranodal lymphoma associated with a chronic immune response to bacteria, viruses, or the autoimmune system. The optimal management of MALT lymphoma has not been clearly established[9]; therefore, several treatment options, such as RT, surgery, chemotherapy alone, and a combination of these modalities, are chosen according to the individual case.[9] RT could be a good treatment option, as it is highly sensitive to radiation. However, surgical resection is used as the initial therapy for nongastric MALT lymphoma when the tumor is localized or not amenable to RT.[9] Patients with nongastric MALT lymphoma have a favorable outcome with a 5-year overall survival rate of 86%.[2] However, MALT lymphoma is a heterogeneous disease and has the potential to progress to a high-grade histologic subtype, such as diffuse large B-cell lymphoma. Regular monitoring and follow-up are important for detecting changes in the disease over time.[10]

The preoperative pathological diagnosis could have impacted the treatment strategy for MALT lymphoma and prostate cancer in this case. However, surgical resection of the urachal tumor was a minimally invasive and effective approach, providing an accurate pathological diagnosis and curative treatment. Although evidence is limited, total resection of the tumor and the urachal ligament, umbilicus, and bladder dome is the mainstay in the management of localized urachal carcinoma.[7] When the surgery is technically feasible, the resection of a localized urachal tumor suspected of malignancy is effective.

Conclusion

Here, we present a case of MALT lymphoma that originated in the urachus. Due to their infrequency, preoperative diagnosis of urachal tumors is challenging. In cases of localized diseases, surgical excision of the tumor can be beneficial for both pathological confirmation and effective disease control.

Author contributions

Kazuma Tsuboi: Writing – original draft. Kensuke Bekku: Writing – review and editing. Kohei Haisa: Writing – review and editing. Yuta Kajihara: Writing – review and editing. Takuji Tsugawa: Writing – review and editing. Yosuke Inoue: Writing – review and editing. Wataru Murao: Writing – review and editing. Tomoko Sako: Writing – review and editing. Shin Ebara: Writing – review and editing.

Conflict of interest

The authors declare no conflict of interest.

Approval of the research protocol by an Institutional Review Board

Not applicable.

Informed consent

Not applicable.

Registry and Registration No. of the study/trial

Not applicable.

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