A case of complete response to chemotherapy followed by cystectomy for adult-onset rhabdomyosarcoma of the bladder

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ABSTRACT

We report a case of alveolar rhabdomyosarcoma of the bladder which achieved complete response with neoadjuvant chemotherapy and surgery. A 58-year-old man was referred to our hospital due to bladder tumor discovered with urinary frequency and gross hematuria. Pathological diagnosis was alveolar rhabdomyosarcoma arising in the bladder which was an extremely rare type. Because the images showed no metastasis except for the right external iliac lymph node, curative treatment was performed consisting of cystectomy besides neoadjuvant chemotherapy using adriamycin and ifosfamide. No residual tumor was found in the extracted bladder specimen. He has been disease free at 12-month follow-up.

1. Introduction

Rhabdomyosarcoma is the most common soft tissue sarcoma in children. Approximately 15%–20% of all cases of RMS are of genitourinary origin. However, RMS of the bladder rarely occurs in adults. Only a few adult cases were reported in the literature. Here, a rare case of bladder RMS in adult male who was successfully managed with neoadjuvant chemotherapy and surgery has been reported.

2. Case presentation

This is the case of a 58-year-old man with no specific medical and family history, who visited a previous doctor due to frequent urination and gross hematuria. At the clinic, a bladder tumor was found, and the patient was referred to our hospital for a thorough examination. Laboratory data at initial examination showed anemia (hemoglobin, 10.3g/dl), and abdominal computed tomography (CT) and magnetic resonance image (MRI) showed a mass on the right wall of the bladder without hydronephrosis. On the 3rd day after the first visit to our hospital, bladder tamponade occurred, and he was admitted to an emergency. A transurethral resection of bladder tumor (TURBT) was performed for hemostatic and biopsy. Cystoscopy results showed a smooth-surfaced, broad-based, nonpapillary tumor on the right wall of the bladder. Pathological results showed diffuse proliferation of large atypical cells with a high N/C ratio, destroying the intrinsic muscularis propria. Since the epithelium was preserved, it was unlikely to be a common urothelial carcinoma, and immunohistochemical studies were added to the diagnosis. Immunohistochemical analysis showed MyoD1 positivity, Desmin positivity, CD56 positivity, and synaptophysin positivity, which are specific for rhabdomyosarcoma (Fig. 1). Conversely, the other immunohistochemical stains, CK7 and CK20 for urothelial carcinoma, CAM5.2 for small cell carcinoma, oSMA for leiomyosarcoma, S100 and HMB45 for nervous system tumor, LCA and myeloperoxidase for malignant lymphoma, and CD34 for angiosarcoma, all yielded negative results. The tumor was a monotonous proliferation of immature tumor cells with a focal structure. There was no myxomatous substrate characteristic of the embryonal type. The final pathological diagnosis was alveolar RMS, with no urothelial carcinoma component.

At the initial visit, CT and MRI showed a mass on the right wall of the bladder, but no lymph node metastasis. (Fig. 2A). CT scan immediately followed by TURBT showed an enlarged bladder mass and the appearance of right external iliac lymph node metastasis, although no distant metastases were observed (Fig. 2B). Based on the histological and radiological diagnosis, we decided that completing surgery was difficult at this status according to the guideline for soft tissue tumors. Therefore, neoadjuvant chemotherapy (NAC) was applied to the first-line radical treatment. For the regimen of chemotherapy, adriamycin plus ifosfamide (AI), was chosen a regimen similar to that used for unresectable advanced or recurrent malignant soft tissue tumors. CT and MRI images post three cycles of AI showed complete response (CR) in both

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bladder tumor and the right external iliac lymph node, and no adverse events occurred during NAC (Fig. 2C). Thereafter, radical cystectomy with pelvic lymph node dissection and ureterostomy were performed. The pathological results showed no residual tumor in the lymph nodes, nor in the primary bladder (pCR) (Fig. 3). The patient has been disease free 12 months post surgery.

3. Discussion

RMS is a malignant tumor that arises from a normal skeletal muscle cell. RMS in the urinary bladder has been well documented in children and adolescents, however in adult, only about 50 cases were reported so far. The majority of RMS embryonal type, with alveolar type RMS, is rare. To the best of our knowledge, only a few adult cases of RMS in the urinary bladder have been reported in literature. The average age of reported adult-onset cases was 63 ± 13 years. A male predominance was observed with a male-to-female ratio of roughly 2 to 1. The tumor usually arises in the trigone and invades the surrounding tissue, presenting as a painful or painless mass. The tumor recurs frequently and metastasizes to the regional lymph nodes, lungs, or liver. The main symptoms of the disease include hematuria, dysuria, and, more generally, bladder dysfunction.

RMS in adult urinary bladders has been reported to be embryonal, alveolar, pleomorphic, or unspecified type. Staining of cells with anti-myogenin or anti-MyoD1 antibodies helps in the differential diagnosis. Additionally, RMS may be positive for the neurological markers synaptophysin and chromogranin. In our case, pathological examination showed alveolar RMS consisting of strongly eosinophilic round cells with diffuse strong desmin expression and strong diffuse vimentin expression and nuclear staining for MyoD1. The neuroendocrine markers CD56, synaptophysin, and chromogranin were also positive.

For adult bladder RMS, treatments vary, including surgical resection, radiotherapy, chemotherapy, or combined therapy. Previous reports have shown good prognosis for resectable cases, but poor prognosis for chemotherapy for recurrence and metastasis.

Moreover, as a pathologic differentiation of bladder tumor malignancies, sarcomatoid carcinoma and the sarcoma in this case are different disease concepts. Sarcomatoid carcinoma is a mixture of urothelial carcinoma and sarcoma components, whereas sarcoma is a pure sarcoma without urothelial carcinoma components. Sarcomatoid carcinoma treatment is generally gemcitabine plus cisplatin chemotherapy, a regimen similar to urothelial carcinoma, whereas sarcoma is treated with VAC (vincristine, actinomycin D, cyclophosphamide) or AI, regimens similar to soft-tissue sarcoma, and the chemotherapy regimens are different. We chose AI therapy in the hope of tumor shrinkage after seeing reports of efficacy with AI therapy.

According to the guideline, this case showed no distant metastasis but lymph node metastasis, so chemotherapy was administered. After chemotherapy, the tumor became resectable, and a cystectomy was performed as a curative therapy, and since the resection margins were negative, the patient is still under observation and has not recurred one year after the surgery.
Given the good clinical outcomes in our case, a complete surgical excision with NAC might be considered one of the treatment options in this type of malignancy.

4. Conclusion

This case is the report on the successful management of a rare alveolar type RMS of the bladder with NAC and cystectomy.

Consent

Written consent to publish was obtained from the patient for the publication of this case and any accompanying images.

Author contributions

Ayato Ito: Conceptualization; data curation; investigation; methodology; project administration; writing – original draft. Kie Sekiguchi: Project administration. Tomohiko Matsuura: Project administration. Yoichiro Kato: Project administration. Masamichi Suzuki: Investigation; project administration. Wataru Obara: Supervision; writing – review and editing.

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Declaration of competing interest

The authors have no conflicts of interest to declare.

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References

1. Shapiro E, Strother D. Pediatric genitourinary rhabdomyosarcoma. J Urol. 1992;148:1761–1768. https://doi.org/10.1016/S0022-5347(17)37023-4 ([PubMed] [CrossRef] [Google Scholar]).
2. Japanese Orthopaedic Association (JOA). Clinical Practice Guidelines on the Management of Soft Tissue Tumors. 2020 ([PubMed]).
3. Bing Z, Zhang PJ. Adult urinary bladder tumors with rabdomyosarcomatous differentiation: clinical, pathological and immunohistochemical studies. Diagn Pathol. 2011;6:66. https://doi.org/10.1186/1746-1596-6-66 ([PMC free article] [PubMed] [CrossRef] [Google Scholar]).
4. Little DJ, Ballo MT, Zagars GK, et al. Adult rhabdomyosarcoma. Outcome following multimodality treatment. Cancer. 2002;95:377–388.
5. Sandler E, Lyden E, Ruymann F, et al. Efficacy of ifosfamide and doxorubicin given as a phase II “window” in children with newly diagnosed metastatic rhabdomyosarcoma: a report from the Intergroup Rhabdomyosarcoma Study Group. Med Pediatr Oncol. 2001;37:442–448.