1. Introduction

Alveolar soft part sarcoma (ASPS) is an exceptionally rare disease among soft tissue sarcoma cases [1]. Only four adult cases of primary ASPS of the bladder have been reported [2–5], and there is no reported pediatric case to the best of our knowledge. We report here our experience with a pediatric case of primary ASPS of the bladder.
was indicated due to the possibility of residual tumor in the bladder. For urinary diversion, construction of a continent urinary reservoir for self-catheterization (Mainz pouch technique) and the abdominal (umbilical) continent catheterizable stoma using the appendix were selected. No residual tumor was observed macroscopically or histopathologically in the removed bladder. The patient’s clinical course after surgery has been favorable. She performs clean intermittent self-catheterization using the abdominal (umbilical) continent catheterizable stoma six times per day, and to date, no urinary incontinence or stricture of the catheterization route has been observed. The patient is under the postoperative follow-up observation with triannual contrast-enhanced CT and annual PET scan. To date, for 2.5 years postoperatively, the patient has been free of any local recurrence or distant metastasis of sarcoma.
Figure 2: (a) Alveolar-structured intense proliferation of tumor cells with a large acidophilic cytoplasm accompanied by abundant capillary vessels is observed with hematoxylin and eosin (H&E) staining. (b) The tumor cells are positive for periodic acid-Schiff (PAS) staining. (c) The tumor cells with pachychromatic nuclei are observed with TFE3 staining.

3. Discussion

ASPS is an exceptionally rare disease accounting for only 0.5-1% of soft tissue sarcoma cases, with the lower extremity being a common site of the primary [6]. The most frequent age of onset is 15-35 years, and it is reported as relatively common in women [6]. Progression of ASPS is generally slower than other types of soft tissue sarcoma and the prognosis has been considered relatively favorable but the cases with distant metastasis are unfavorable. According to a previous report, survival rates of 2, 5, 10, and 20 years are 87%, 62%, 43%, and 18%, respectively [6].

As the primary local lesion is often asymptomatic, ASPS is likely to be detected owing to the subjective symptoms associated with a metastatic lesion, and the prognosis of such cases with distant metastasis is usually poor [6]. Distant metastasis is most likely to occur in the lung (38%), the bone (33%), and the brain (32%), and the mean survival duration after metastasis development is 2 years [6].

The present article is the first report of a pediatric case of primary ASPS of the bladder. After this patient was histopathologically diagnosed with ASPS based on the results from TUR-Bt, cystourethrectomy was performed to completely remove the tumor due to the possibility of residual tumor in the bladder. The progress of the patient has been favorable for 2.5 years postoperatively, without any recurrence or metastasis.

Table 1 summarizes the four adult cases of primary ASPS of the bladder previously reported. As with our case, all adult cases were detected by the development of urinary symptoms. Surgical treatment was selected for three of the cases, but treatment details were unspecified for case 2. Regarding distant metastasis, the lesions for cases 3 and 4 were locally restricted in the bladder and no distant metastasis or recurrence occurred during the follow-up period; details are unknown for cases 1 and 2. Local symptoms of primary ASPS of the bladder, such as urinary symptoms (e.g., macroscopic hematuria), develop at an earlier stage than primary ASPS of other organs. In addition, ASPS has a slowly progressive nature [6]. Therefore, we assumed that early detection and treatment may be possible before development of the distant metastasis. As residual tumor was not histopathologically observed in the removed bladder, cystourethrectomy may have been deemed overtreatment. For ASPS with residual tumor, complete resection of the tumor improves the patient’s prognosis [7].
As shown in Table 1, there has been no reported treatment option other than surgery. Considering curability as the first priority, we believe that cystourethrectomy was an appropriate treatment strategy in this case.

The first-line choice for treating primary ASPS of other organs is also surgery if no distant metastasis has developed and resection of the primary lesion is applicable [6]. According to a previous report, treatment options for ASPS patients for whom surgery is not applicable or with distant metastasis or postoperative residual tumor include monotherapy or combination therapy of anticancer agents, molecular target agents, and radiation therapy. However, efficacy of those treatment options has not been clearly demonstrated [6–9].

The present case showed the alveolar-structured intense proliferation of tumor cells with a large acidophilic cytoplasm accompanied by abundant capillary vessels. Also, the patient's TFE3 results were positive, indicating a histopathological feature of ASPS. The differential diagnosis of ASPS includes carcinoma and melanoma, but lack of definite staining for epithelial and melanoma markers rules out these possibilities. Granular cell tumor may exhibit morphologies similar to epithelial and melanoma markers rules out these possibilities. However, efficacy of those treatment options has not been clearly demonstrated [6–9].

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Table 1: Four adult cases of primary bladder ASPS previously reported.

| patient | age | sex | chief complaint | treatment | local recurrence | metastases | follow-up | outcome |
|---------|-----|-----|----------------|-----------|-----------------|------------|-----------|---------|
| case 1 [2] | 31 | male | hematuria, urinary frequency | partial cystectomy | + | unknown | 17 months | death |
| case 2 [3] | 37 | male | narrow urinary stream | unknown | unknown | unknown | unknown | unknown |
| case 3 [4] | 25 | female | hematuria, dysuria | TUR-Bt | – | – | 45 months | alive |
| case 4 [5] | 18 | male | macroscopic hematuria, dysuria | TUR-Bt, partial cystectomy | – | – | 28 months | alive |

TUR-Bt, transurethral resection of the bladder tumor.

4. Conclusion

Complete resection of tumor is the only definitive treatment for ASPS. The efficacy of molecular target agents, anticancer agents, and radiation therapy has not been clearly demonstrated. Although the residual tumor was not observed in the removed bladder or urethra in the present case, we believe that cystourethrectomy was an appropriate treatment because the prognosis would have been poor if the residual tumor had been present. In pediatric patients presenting with asymptomatic macroscopic hematuria, the possibility of primary ASPS of the bladder should be considered, even though the incidence may be low.

Since the course is indolent with 18% survival at 20 years, strict follow-up is mandatory. We might consider opting a follow-up with cystoscopy or a second TUR in similar case.

Conflicts of Interest

All authors declare that they have no conflicts of interest.

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