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Transurethral resection for botryoid bladder rhabdomyosarcoma

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ABSTRACT

The outcome of multimodal therapy for localized bladder rhabdomyosarcoma is quite good in terms of morbidity, and conservative surgery is generally recommended. However, in cases originating in the bladder neck, tumorectomy or partial cystectomy has adverse effects on bladder function. A 2-year-old girl underwent transurethral resection of a bladder tumor (TUR-BT), chemotherapy consisting of vincristine, actinomycin-D, and cyclophosphamide, and radiotherapy. She was in remission for 3 years when frequent urination became evident. Her bladder capacity and compliance were low; however, her urinary symptom was controlled using anticholinergic medication. Accordingly, TUR-BT could be an optional approach for bladder rhabdomyosarcoma.

1. Introduction

Rhabdomyosarcoma is the most common pediatric soft tissue sarcoma. Approximately 10–15% of all rhabdomyosarcoma cases arise from the pelvic organs [1]. The tumor is often large and involves adjacent organs and vessels. Histologically, bladder/prostate rhabdomyosarcoma is typically either embryonal or botryoid.

After chemotherapy, radical surgery is required; this is usually urinary reconstruction or diversion, except for tumors arising at the top of the bladder [2]. Recently, multimodal treatment with chemoradiotherapy and surgery has led to a good prognosis. Therefore, conservative surgery (subtotal/partial resection) is generally recommended [2]. Laparotomy is often used for bladder/prostate rhabdomyosarcoma; however, transurethral resection of a bladder tumor (TUR-BT) can be applied to preserve the organ when the tumor is growing into the bladder, provided that the residual disease can be treated with chemoradiotherapy. Herein, we describe a patient with bladder rhabdomyosarcoma that was resected using TUR-BT.

2. Case report

A 23-month-old girl presented with macroscopic hematuria and a vulval mass. At admission, enhanced computed tomography (CT) indicated a solid mass measuring 32 × 30 × 26 mm that occupied the bladder (Fig. 1A and B) without metastasis. A biopsy was performed using cystoscopy and showed increased numbers of rhabdomyoblasts and polygonal-shaped cells with eosinophilic cytoplasm (Fig. 1G and H). She was diagnosed with botryoid embryonal rhabdomyosarcoma, group III, stage 2, and vincristine, actinomycin-D, and cyclophosphamide (VAC) therapy was administered [3]. The tumor shrank (Fig. 1C and D), and TUR-BT was performed after 8 weeks. The tumor was localized in the bladder

Abbreviations: TUR-BT, Transurethral resection of a bladder tumor; CT, Computed tomography; IRS, Intergroup Rhabdomyosarcoma Study; SIOP, International Society of Pediatric Oncology; MMT, Malignant Mesenchymal Tumors.

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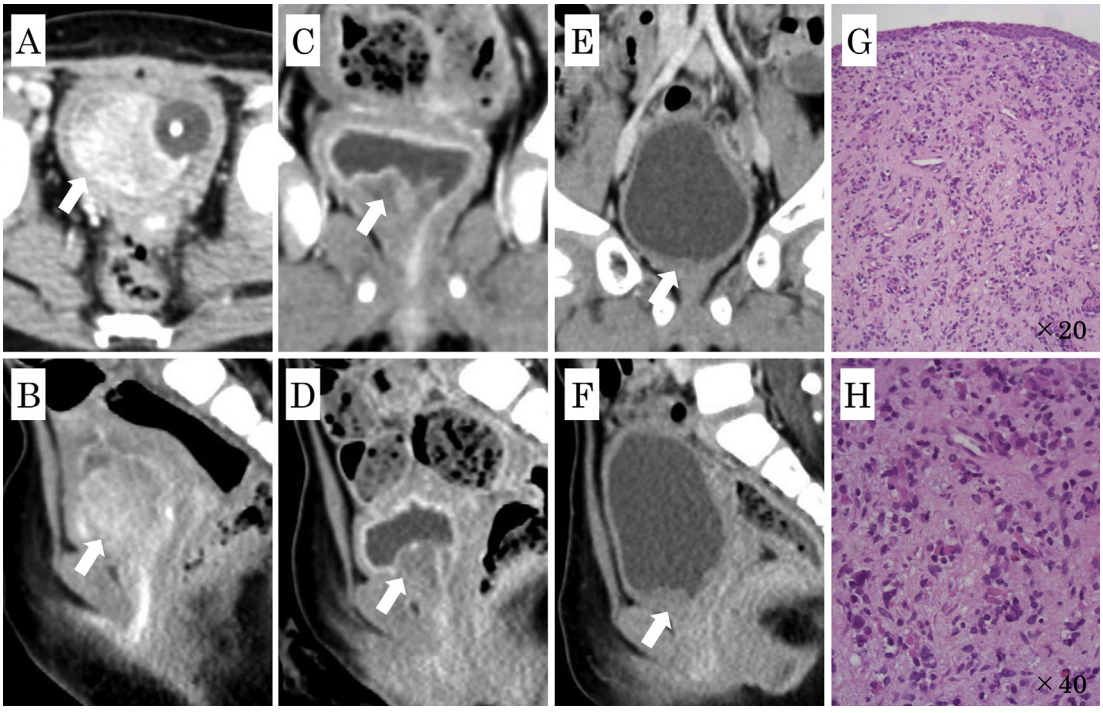

Fig. 1. Computed tomography and histopathological findings. A, B: An enhanced solid mass measuring 32 × 30 × 26 mm occupying the bladder (white arrow). C, D: An enhanced mass remained in the bladder neck (white arrow). E, F: Wall thickening of the bladder neck after chemoradiotherapy (white arrow). G, H: Rhabdomyoblasts and polygonal-shaped cells with eosinophilic cytoplasm increased pathologically.

neck and was resected while checking with echo, except for the muscle layer of the bladder neck. After surgery, chemotherapy and radiotherapy was administered. The proton beam was used to irradiate the residual tumor site at 50.4 Gy (RBE: relative biological effectiveness) and the entire bladder was irradiated with 30.6 Gy (RBE). At the end of therapy, enhanced CT indicated that the bladder neck wall was still thickened (Fig. 1E and F). Therefore, an additional biopsy was transurethrally performed, which showed that this thickened lesion consisted of fibrotic tissue that did not contain viable cells. There has been no relapse of the tumor 3 years after the end of treatment.

After discharge, the patient developed persistent increased urinary frequency and transient urinary hemorrhage. Two years after therapy, voiding cystography indicated that the bladder capacity was 84 mL (56%) at 4 years of age (normal level 150 mL) (Fig. 2A) [4] and the bladder neck wall was irregular. A urodynamics study indicated an intravesical pressure of 16 cmH_2O at the maximum bladder volume of 83 mL (Fig. 2B). This resulted in a poorly compliant bladder. However, the symptom was controlled using anticholinergic medication and restriction of fluid intake.

3. Discussion

The appropriate treatment strategy for bladder/prostate rhabdomyosarcoma is combined modality treatment with chemoradiotherapy and surgery. The prognosis of localized embryonal bladder/prostate rhabdomyosarcoma is good [5], and conservative therapy improves patients' quality of life, as demonstrated by clinical trials [6]. However, organ preservation in the surgical process remains controversial. Conservative surgery includes tumorectomy, partial prostatectomy, partial cystectomy, or both partial prostatectomy and cystectomy, in which the posterior urethra is spared. Radical surgery is typically performed with margin-free tumor resection with concordant removal of the bladder and the posterior urethra, and is associated with lymph node dissection [7]. Filipas et al. stated that conservative surgery carries high risks of positive margins and tumor relapse, and that radical surgery and current techniques of continent urinary diversion are beneficial [2]. However, Alexander et al. showed no difference between conservative and radical surgery in terms of the 5-year overall and event-free survival of patients with bladder/prostate rhabdomyosarcomas [8]. In the Intergroup Rhabdomyosarcoma Study (IRS) IV study, the overall survival rate of patients with nonmetastatic bladder/prostate rhabdomyosarcoma was 82% (72/88). Fifteen of 88 patients needed urinary diversion [6]. In the International Society of Pediatric Oncology (SIOP) study, the overall 5-year survival rate of patients with nonmetastatic bladder/prostate rhabdomyosarcoma was 73% [7]. In both studies, organ preservation surgery was used.

Cystoscopy for bladder rhabdomyosarcoma is usually performed only for biopsy or exploration, and not for definitive surgery, because the rhabdomyosarcoma arises from the muscle layer of the bladder. TUR-BT was performed for transitional cell carcinoma in children, as well as in adults [9,10]. Complications of this procedure include postoperative bleeding and perforation of the bladder wall. In particular, surgeons must be careful of excessive resection of tumors arising from the bladder wall. Although potentially carrying the risk of residual disease, TUR-BT may be sufficient for local control of bladder rhabdomyosarcoma, when patients are properly treated with postoperative radiotherapy.

It is speculated that the cause of incontinence after conservative surgical therapy for bladder/prostate rhabdomyosarcoma is the origin of a tumor or tissue damage due to radiotherapy. For conservative surgery, radiotherapy with consideration of both curability and tissue damage mitigation is an important factor. Proton beams and brachytherapy have been reported to be useful methods for reducing tissue damage by radiation therapy. For bladder/prostate primary tumors, when there is a residual tumor, 50.4 Gy (RBE) of external irradiation is required; however,