



Case report

Abol-Enein pouch modification after radical cystectomy in bladder rhabdomyosarcoma in 5 years old child during COVID-19 pandemic: A case report

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ABSTRACT

Objective: To discuss the consideration of performing radical cystectomy for rhabdomyosarcoma in children during the COVID-19 pandemic in the urology department of Adam Malik Hospital, Medan.

Introduction: Rhabdomyosarcoma is a rare malignancy that develops from primitive mesenchymal stem cells. The gold standard for the treatment of rhabdomyosarcoma is radical cystectomy. However, during the COVID-19 pandemic, radical cystectomy becomes a concern due to the risk of virus transmission. This article reported a patient who underwent radical cystectomy during the COVID-19 pandemic.

Case presentation: A five-year-old female child was admitted to the hospital with chief complaints of bloody urine 1 month before admission. A month before hospital admission, the patient had pain during urination and was treated with radical cystectomy with Abol-Enein pouch modification.

Result: After the surgery, the patient was able to urinate without any disturbance. Hematuria was not found. There was no complaint related to stricture of the ureter after the surgery.

Conclusion: In our center, radical cystectomy could be performed in child with bladder rhabdomyosarcoma during COVID-19 pandemic. The procedure is considered a high priority therapy in most regions during COVID-19 pandemic. Abol-Enein technique after radical cystectomy resulted in improvement of symptom with no reported complication in our case.

1. Introduction

Rhabdomyosarcoma (RMS) accounts for the majority of pediatric soft-tissue sarcoma. The tumor develops from primitive mesenchymal cells which should undergo differentiation into skeletal muscle [1]. The distribution of cases is high during first decade and adolescence. Rhabdomyosarcoma is diagnosed in 4,5 cases/million children/adolescents every year [2]. Urogenital system rhabdomyosarcoma was found in 15–20% of all RMS [3].

The age distribution of RMS follows bimodal distribution with the first peak occurs in the age of 0–5 years and the second smaller peak occurs in adolescence. There are two major subtypes of RMS which are embryonal (ERMS) and alveolar rhabdomyosarcoma (ARMS). ERMS is diagnosed earlier before 10 years of age with peak incidence between

0 and 4 years of age (42%) while ARMS is diagnosed equally in 0–19 years of age [4]. Between the year of 1975 and 2005, rhabdomyosarcoma were diagnosed in 987 children aged 0–19 years. The majority of subtypes found were ERMS (57%) and ARMS (23%). Other less common subtypes found were embryonal sarcoma (2%), pleomorphic (1,5%), mixed type (1,4%), and spindle cell (0,6%) [4].

Radical cystectomy is a gold standard for therapy of rhabdomyosarcoma. The timing of the procedure determined the prognosis and survival of patients with rhabdomyosarcoma [5]. According to Sharma, radical cystectomy in high risk cancer is a procedure not to be cancelled during COVID-19 pandemic while radical cystectomy in low risk cancer is optional or secondary to cancellation [6]. BAUS guidelines reported that low risk patient needing radical cystectomy could be in secondary cancellation while high risk patient is last to be cancelled [7]. During

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COVID-19 pandemic, radical cystectomy had been performed in several center in several countries such as Turkey and Europe.

Following radical cystectomy, bladder substitution is conducted. Abol-Enein and Ghoneim performed an implantation of ureters into a detubularized bowel segment reservoir. The ureters were placed between the layer of tubularizing folds so that the serous-lined extramural tunnel was formed. The intraluminal pressure prevent reflux. The procedure could be performed for normal size ureter and grossly dilated and thick-walled ureter. Besides, the alternative method for neobladder creation was the I-pouch. However, the using of the procedure in a pediatric population has not been reported [8]. This article report a female patient, 5 years old with rhabdomyosarcoma of the bladder who underwent radical cystectomy. This case report was written according to SCARE guideline [9].

2. Case presentation

A female child 5 years of age admitted to the hospital with chief complaint of blood in urine 1 month before admission. There was loss of weight up to 5 kg in the last 1 month. The patient was diagnosed with bladder rhabdomyosarcoma T3N3M0. On the laboratory examination, the hemoglobin was 10,6 g/dl, leukocytes were 9300 cells/ μ l and thrombocytes was 628.000 cells/ μ l. Serum ureum was 32 mg/dl and serum creatinine was 0.42 mg/dl. Serum electrolytes were in normal limit. Serum albumin was 3.0 mg/dl. The production of conduit was 11,000 cc/24 h. The patient lived with her parents. Her father was a heavy smoker and her parents often used pesticide in the garden near their house.

The patient underwent radical cystectomy with Abol-Enein pouch modification. The ileal conduit was performed after radical cystectomy. Figs. 1–4 demonstrated the procedure of radical cystectomy with Abol-Enein pouch modification. After the surgery, the pain improved. There was no macroscopic and microscopic hematuria reported. No stenosis of the ureters was found. Figs. 5 and 6 showed the postoperative abdominal physical and radiological examination. The patient was stable and discharged with no complication. 1 month after the surgery, the patient came to the hospital for follow-up examination and no complication was reported.

3. Discussion

Rhabdomyosarcoma is the majority of soft-tissue sarcoma found among children. It accounts for 5% of all childhood cancers [1]. The finding of rhabdomyosarcoma in children is rare [1]. It represents about 3% of all childhood cancers. Rhabdomyosarcoma mostly occurs in children and teens [10]. Most rhabdomyosarcoma (87%) occurs in the patients younger than 15 years [11]. The incidence was slightly higher in male compared with female [10]. The rhabdomyosarcoma was classified into two main types which are embryonal rhabdomyosarcoma



Fig. 1. Ileal segment identification.

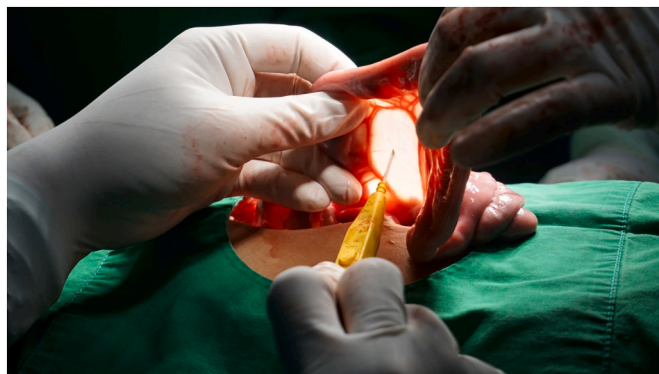


Fig. 2. Ileal segment for Abol-Enein pouch modification.

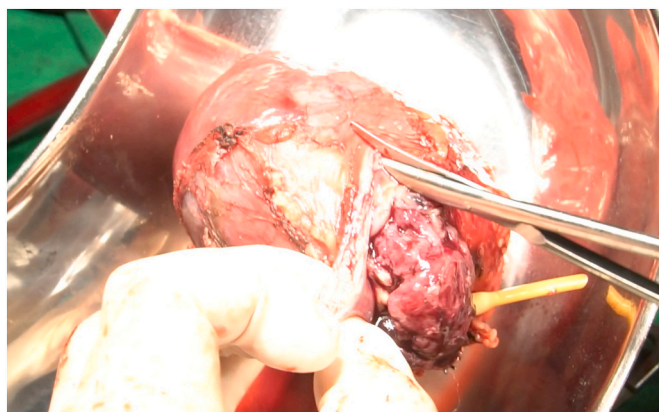


Fig. 3. Bladder tumor identification.

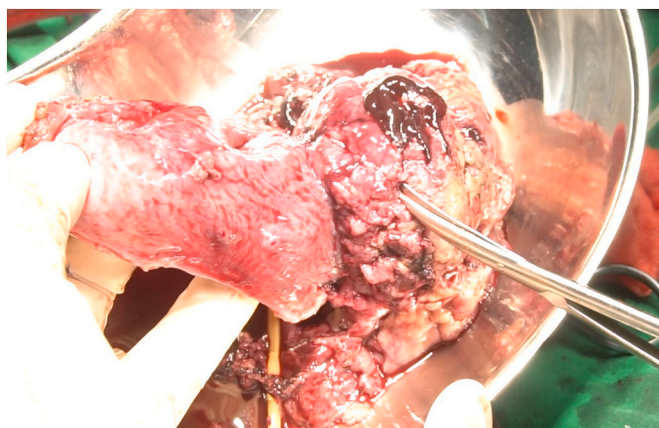


Fig. 4. Bladder tumor after bladder incision.

(ERMS) and alveolar rhabdomyosarcoma (ARMS) [10].

Genitourinary rhabdomyosarcoma represents approximately 25% of all rhabdomyosarcoma [11]. The development of rhabdomyosarcoma is sporadic which could be affected by several risk factors. Several cases of rhabdomyosarcoma had familial predisposition such as Li-Fraumeni syndrome and neurofibromatosis. There are still few studies exploring the relationship between several risk factors including pre-conceptional use of recreational drugs, prenatal X-ray exposure, maternal history of stillbirths, advanced maternal age at childbirth, high birthweight, parity, and the risk of rhabdomyosarcoma in children [12]. Grufferman S et al. stated that fathers' cigarette smoking was associated with childhood rhabdomyosarcoma with relative risk of 3.9 [13]. In this report, we



Fig. 5. Postoperative abdominal physical examination.

found that the patient's father was a heavy smoker. This could be risk factor of childhood rhabdomyosarcoma in our patient.

The common clinical presentation of bladder rhabdomyosarcoma was hematuria. In our patient we found gross hematuria. Priyadarashi also reported that microscopic hematuria was found in bladder rhabdomyosarcoma [11]. Other clinical manifestations that could present include straining during voiding, sense of incomplete voiding, weak stream urine, suprapubic tenderness, and dribbling of urine [11]. Our patient also had pain related to the tumor.

Radical cystectomy remains the gold standard for treatment of bladder cancer. The timing of the procedure is essential since reduction in overall survival and in progression-free survival was observed

following a 90-day delay in conducting radical cystectomy [5]. However, during COVID-19 pandemic, there are several considerations of performing the surgery including increased risk of mortality and morbidity in COVID-19 patients, increased risk of healthcare worker get infected from the patients, and the limitation of facility used which could be reserved in case there will be an increase in COVID-19 patients admitted to inpatient care [14]. Cancer patients are in the immunosuppressive state. During the COVID-19 pandemic, cancer patients have a higher-risk for developing severe manifestation that leads to ICU admission, requirement for mechanical ventilation, and death [15].

There were several reports of radical cystectomy performed in COVID-19 pandemic. Adanur et al. reported that radical cystectomy and ileal conduit operations were performed in eight patients in their center in Turkey. The mean age of the patients was 62,8 years and the patients were diagnosed with muscle invasive bladder tumor between March 11, 2020 and May 22, 2020 [12]. Three patients had wound infection in the post-operative period. The mean hospitalization time of the patients was 11 days followed by discharged with full recovery [12]. Soytaş M et al. also performed radical cystectomy for 2 patients [13]. Gulia et al. stated that the curative surgery for high grade tumor should be performed. The surgery could be delayed in low grade sarcomas. Palliative surgery should be avoided and non-invasive options should be considered. However in COVID-19 positive or highly suspected patients, there could be a delay in performing the procedure until 15 days or until the patients recover from the symptom or have a seronegative result [14].

Gulia et al. stated that primary high-grade sarcoma without metastasis was a priority for surgery [14]. Komarsara L et al. stated that the timing and extent of radical surgery for management of genitourinary rhabdomyosarcoma was determined by local anatomical conditions and conditions following chemotherapy and radiotherapy. The treatment of choice for local therapy of rhabdomyosarcoma is cystectomy continued by reconstructive surgery [15]. According to survey of urologists in Europe, the radical cystectomy was included on the priority list higher than expected rates. The rate of radical cystectomy postponement was lower than expected rates [7]. Overall the priority rank of radical cystectomy in 6 regions (Africa, East/South East Asia, Europe, North America, South America, West/South West Asia) was 8 from the scale of 1 (lowest priority) to 10 (highest priority). Different region had different priority rank of radical cystectomy. Radical cystectomy was ranked 9 in Europe. In Africa, East/South East Asia, North America, and South America, the procedure was ranked 8 while in the West/South West Asia, the procedure was ranked 6 [7]. Carneiro A et al. reported that surgical treatment is recommended for bladder cancer in the COVID-19



Fig. 6. Postoperative abdominal X-Ray examination.

pandemic area since the delay could influence the prognosis. Carneiro A et al. added that the delay of cystectomy should be no more than 10 weeks after chemotherapy. After the cystectomy, adjuvant chemotherapy with cisplatin and gemcitabine is recommended [16].

The technique of ileal conduit with ureteral reimplantation using serous-lined extramural tunnel (Abol-Enein technique) had been reported for children with rhabdomyosarcoma as shown in Fig. 7 [15]. Abol-Enein H et al. performed the uretero-ileal reimplantation technique on 12 patients who underwent cystectomy. They stated that the method of uretero-ileal reimplantation resulting in formation of 2 serous lined extramural tunnel in a detubularized ileal resulted in patent ureter and unidirectional urine flow. The patients were reported to have good continence during the day while 9 of 12 patients were also continent at night without medication. Three patients reported a several degree of nighttime incontinence with good response to imipramine hydrochloride 25 mg which was given at bed time [17]. In addition, Abol-Enein et al. also performed the urinary diversion procedure by using serous lined extramural valve in 18 men and 5 women. The procedure resulted in satisfactory outcome without operative or postoperative mortality. The urinary continence was reported in all patients except 1 patient [18]. Fig. 1 showed the procedure of uretero-ileal reimplantation using Abol-Enein technique [17]. Komasa L et al. stated that all

children with rhabdomyosarcoma which underwent the procedure of radical cystectomy with Abol-Enein technique were all alive and were in complete remission after the treatment [15]. In addition, Komasa et al. reported 1 case of stenosis of distal part of the ureter following the procedure [15]. In our patient, we did not find stenosis of the ureters within the follow-up period of 1 month.

The development of the surgical technique had been reported in previous studies. Torrey RR et al. reported that radical cystectomy with robotic-assisted laparoscopy with Indiana pouch continent cutaneous urinary diversion reconstruction resulted in daytime and nighttime continence except 1 patient. The complication of using laparoscopic technique was comparable with open surgery [18].

4. Conclusion

Radical cystectomy remains the gold standard treatment for bladder rhabdomyosarcoma. The procedure is considered a high priority therapy in most regions during COVID-19 pandemic. In our study, the radical cystectomy was performed in female child patient during COVID-19 pandemic. The postoperative outcome was good without complications. After the surgery, the patient and medical staff were found negative for COVID-19.

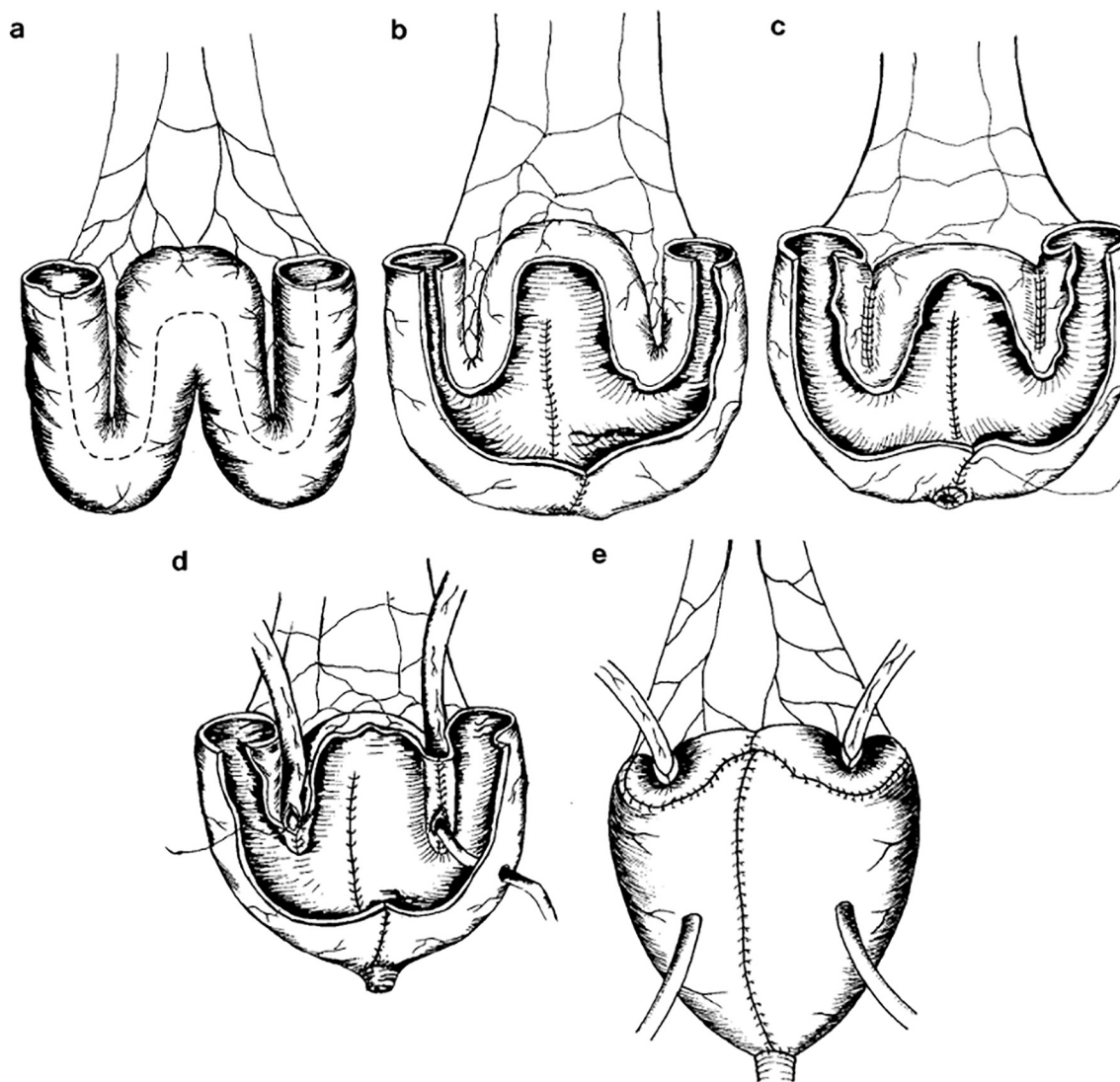


Fig. 7. The procedure of uretero-ileal reimplantation with formation of 2 serous lined extramural tunnels in a detubularized ileal W-bladder. (a) Isolation and arrangement of 40 cm long segment of distal ileum into a W-shaped configuration. (b) Incision of antimesenteric border. (c) The seromuscular suturing of lateral ileal flaps together to form 2 serous lined intestinal troughs (d) Anastomosis of ureter to intestinal mucosa (e) Closure of ileal reservoir following urethra-ileal anastomosis.

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Ethical approval

This case report has been exempted from ethical approval by Universitas Sumatera Utara Ethical Committee.

Consent

Written informed consent was obtained from the patient and her parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Guarantor

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Not applicable.

Availability of data and materials

The datasets generated during and/or analyzed during the current study are available on demand.

CRediT authorship contribution statement

JTG carried out the data collection, analyzing the data and drafted the manuscript. YS participated in the design of the study and helped to draft the manuscript. All authors have read and approved the manuscript.

Declaration of competing interest

The authors declare that they have no competing interests.

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