RHABDOMYOSARCOMA IN A 16-YEAR-OLD BOY: A RARE CASE REPORT

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ABSTRACT

The urinary bladder is a prominent site for rhabdomyosarcoma (RMS), the second most common malignant soft tissue sarcoma in children and adolescents after osteosarcoma. RMS is an uncommon and occasionally fatal malignancy that affects youngsters. Despite extensive treatment plans that combine radiation therapy, dose-intensive combination chemotherapy, and surgery, the prognosis for individuals with metastatic disease is still dismal.

Keywords: Immunohistochemical findings, Rhabdomyosarcoma, Urinary bladder.

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INTRODUCTION

The most prevalent soft-tissue sarcoma in children is called rhabdomyosarcoma (RMS). Rhabdomyosarcomas make up 40% of soft tissue sarcomas, which make up about 7% of all malignancies in children and adolescents. [1] Nonetheless, they are uncommon, making up only 3% to 4% of all paediatric malignancies.

In patients with bladder rhabdomyosarcoma, lower urinary tract symptoms include hematuria, dysuria, frequent urination, and desire to urinate are frequently present. Nonetheless, some conditions might be asymptomatic and discovered by chance 2.

RMS is divided histologically into two primary subgroups: alveolar RMS (ARMS), which only accounts for 20%–30% of cases, and embryonal RMS (ERMS), which accounts for 60%–70% of cases 3. RMS's aetiology is still a mystery.

CASE REPORT

A 16-old boy was referred to our institution with a pre-diagnosis of Malignant neoplasm of bladder and prostate. On history taking, boy who was vitally stable, reported that he felt burning during micturition along with urine obstruction, incomplete emptying of bladder plus weak stream, hematuria few months ago was advised for ultrasound kidney ureter bladder (KUB) showed right ureteric obstruction, thick-walled urinary bladder as well as bladder outflow obstruction.

His digital rectum examination showed stony hard prostate. His computerized tomography (CT) KUB without contrast was done showed thickening of bladder wall in the multiple places including the left anterior and posterior wall; the bladder trigone and right posterior wall causing obstruction of the right ureter with right side hydronephrosis and enlarged edematous right kidney.

There were multiple enlarged lymph nodes in the pelvis. His Cystoscopy plus transurethral resection of a bladder tumor (TURBT) was done. His histopathology specimen taken after surgery was from bladder neck trigone of urinary bladder, median lobe of prostate and deep muscle sites diagnosed as Rhabdomyosarcoma whereas deep muscle sites showed ulcerated mucosa and detrusor muscle.

After TURBT few days back patient felt severe pain left flank. His ultrasound KUB was repeated after surgery whose impression showed diffused circumferential marked wall thickening/ residual tumor resulting in bilateral hydroureteronephrosis more on right side. Bilateral grade 1 renal parenchymal changed.

His MRI was repeated reports were worrisome for malignant neoplasm of urinary bladder and right distal ureter with bilateral pelvic and retroperitoneal lymphadenopathy as the biopsy reported Rhabdomyosarcoma with creatinine 1.52 (Figure 1).

Meanwhile patient received chemotherapy of vincristine, Actinomycin D and cyclophosphamide (VAC) and Ifosfamide and etoposide (IE).

After that PET CT scan was done to evaluate the response of chemotherapy and it was also compared and correlated with previous PET CT and MRI pelvis. Results

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showed that residual bladder thickening along the left posterolateral aspect where metabolic activity cannot be commented upon due to physiologic excretion of radiotracer through bladder. A few non-enlarged bilateral pelvic sidewall nodes were metabolically insignificantt. After few days, his MRI was repeated to compare with the last MRI as the patient had no active complain.

This time MRI report concluded as unremarkable appearance of urinary bladder with no measurable disease as well as no size significant pelvic lymph nodes observed. Meanwhile two cycles of chemotherapy were repeated. Now his MRI was repeated from our institution and is compared with previous investigations. Results concluded no evidence of any significant bladder wall thickening.

Bladder rhabdomyosarcomas are rare mesenchymal malignancies in children with a poor prognosis. Here, in our case, surgical treatment like cystectomy and radiotherapy is not applicable for treatment for localized forms. Chemotherapy was done in past 8 months as mentioned earlier in which of seven cycles of vincristine, Actinomycin D and cyclophosphamide (VAC) and six cycles of Ifosfamide and etoposide (IE) with gap of two weeks. The role of chemotherapy in the management of metastatic forms worked.

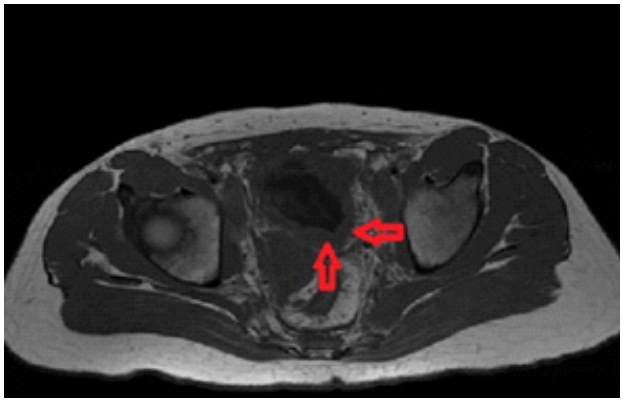

Figure 1: MRI Pelvis with contrast
(Red arrows show bladder wall growth)

DISCUSSION

Children aged 1-4 years have the highest incidence of RMS, followed by those aged 10–14 years and 15–19 years with the lowest frequency. Our patient was a 16-year-old boy who was quite high-risk. These tumours grow rapidly and aggressively, enlarge rapidly, are typically painless, and are associated with high recurrence rates and widespread metastases via hematogenic and/or lymphatic pathways. [4] Thankfully, in this instance, the patient's guardians' cooperation, timely consultation for adjuvant treatment, and follow-up at our institution may have favored the tumor's quick advancement and care and, as a result, lessened the severity of the disease. Paediatric patients with bladder and prostate embryonal rhabdomyosarcoma have a worse prognosis than those with the illness in the vagina, uterus, or Para testicular area (respectively, 70%-73% and 84%-89%). [5] Due to its rarity, the prognosis and ideal care of adult embryonal rhabdomyosarcoma are unknown.

There is no set course of action providing for improved survival. Although several treatments have been suggested, it appears that a multimodal approach is required. Transurethral bladder resection, chemotherapy, radiation, and total or partial cystectomy are among the available treatments. [6,7]

We chose chemotherapy following TURB since, in our situation, RMS was shown to be metastatic, which allowed the haematuria to stop. The best treatment option for localised RMS is still radical cystectomy combined with bilateral lymph node dissection. Neo-adjuvant chemotherapy may be performed before this major operation. In cases of embryonal RMS of the bladder in children, radiation therapy used in conjunction with surgery produces great results. When the tumour is tiny (less than 3 cm) and situated at the level of the dome, partial cystectomy can be suggested for young men who wish to father children. [8]

CONCLUSIONS

A multidisciplinary strategy is necessary to overcome advance staging to provide timely therapy. As RMS tends to spread to the bone marrow, it entails surgical excision of the tumour followed by multi-agent chemotherapy with or without radiotherapy. Our case is the first one we are aware of that successfully managed RMS of the urinary bladder by chemotherapy and surgery. Nonetheless, regular follow-up and patient compliance also contribute to the success of the treatment.

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