Oncology

Prostatic rhabdomyosarcoma revealed by acute urinary retention

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ABSTRACT

Rhabdomyosarcoma of the urinary tract is very rare among young adult and the survival has improved significantly. The challenge of such location is to ensure better functional outcomes like continence and sexuality without compromising the overall survival.

Throughout this case, we bring new insights into the conservative management of prostatic sarcoma and we focus on the role of radio-chemotherapy for local disease control.

Introduction

Rhabdomyosarcoma of the urinary tract is an uncommon tumor. Although cases are sporadic, some genetic factors have been associated with these tumours like neurofibromatosis type I, costello syndrome and Beckwith Widemann syndrome.

The management of genitourinary sarcoma has evolved during the last years, reducing surgical radical approach to ensure better quality of life without compromising survival rates.

We reported a case of embryonal rhabdomyosarcoma arising from the prostate of 17-year-old children.

Case summary

A 17-year-old boy presented with a history of bladder outlet obstruction, initial haematuria for two weeks and then followed by acute urinary retention. He was also complaining of increased frequency and urgency of micturition. The patient reported a feeling of incomplete evacuation and dribbling, during the last 3 months.

He had no fever but a weight loss of 5 Kg. On the physical examination, he had not lymphadenopathy or hepatosplenomegaly. Bone palpation was without pain.

The digital rectal examination revealed a 70 g prostatomegaly with a large growth firm to hard in consistency and irregular in shape. There was no gross rectal involvement.

After performing a transurethral resection (Fig. 1), the histologic exam revealed small egg-shaped tumor cells on hematoxylin and eosin staining. The proliferation is characterized by alternating of cellular and myxoid areas. Mitotic figures were numerous. Myogenin and desmin expression was positive on immunohistochemical examination (Fig. 2).

MRI showed a high-intensity tumor on T2-weighted and diffusion weighted images, which was heterogeneously enhanced on gadolinium-enhanced imaging (Fig. 3a).

The diagnosis was a localized embryonal rhabdomyosarcoma of the prostate, with gross residual disease after incomplete removal, which was classified into stage II group IIIb by the IRS Group staging system. The patient was been staged at an intermediate risk category.

Systemic chemotherapy had been started according to the IRS III VAC regimen, which consisted of 16 weeks combinations of vincristin, actinomycin D and cyclophosphamide. After adjuvant chemotherapy, the patient achieved a complete remission.

Local relapse had been detected within 7 months. The patient did not accept to undergo radical cystoprostatectomy. Then, salvage treatment with radiotherapy was indicated.

A thoraco-abdominal computed tomography (CT) scan after 25 months of follow-up revealed local advanced disease but without metastasis (Fig. 3b).

Discussion

Malignant prostatic tumours of mesenchymal origin are rare, corresponding to 0.3%–1.0% of all prostatic tumours.

Rhabdomyosarcoma, arising from immature mesenchymal cells with skeletal muscle differentiation, is the most frequent paediatric soft tissue sarcoma. But only 15–20% of all rhabdomyosarcoma arise from the urogenital tract.
This tumor is characterized by rapid growth with the formation of pelvic masses. Symptoms are often due to obstruction of the urethra like frequency, hesitancy, dysuria and less commonly haematuria and acute urinary retention.

The presence of constipation and rectal fullness is associated with locoregional involvement.

Classically, three histological subtypes have been identified: Embryonal, pleomorphic and alveolar histiotypes. The embryonal subtype is the most frequent and often associated with favorable prognosis. The alveolar and the pleomorphic variants are less frequent and more aggressive. The alveolar subtype is characterized with by a specific translocation t(2; 13) (q35; q14).

Lactate dehydrogenase and serum prostate-specific antigen (PSA) levels are usually normal due to the non-epithelial origin of prostate sarcoma.

Both Ct scan and MRI could be useful for the positive diagnosis, the regional extension and for the follow-up.

On Ct scan, the tumor has no specific features and appears as an irregular mass filling the pelvis with heterogeneous enhancement after PDCI. This process usually show aggressive behavior with invasion of the bladder wall and rectal compression.

On MRI, Rhabdomyosarcoma usually appears slightly hypointensive on T1-weighted images and hyperintensive on T2-weighted images with internal irregular liquid intense areas.

The presence of metastatic disease at diagnosis is a poor predictor of outcome.

The primary chemotherapy approach for patient with localized disease is the preservation of important structures especially for children without diminishing the likelihood of survival. This is consisting of four to six courses VAC, which is the mainstay of chemotherapy in IRS trials. Although, the German group reported a higher rate of responders (two-thirds or more in terms of tumor volume reduction) in the ifosfamide than the cyclophosphamide-based trial, But, no significant advantage in terms of overall-survival was noticed.

In case of persistence of the tumor after a multi agent chemotherapy, local radiation and/or bladder-sparing surgery is needed.

Radiotherapy had proven good results in multiples studies as a local}

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**Fig. 1.** Transurethral resection of the prostatic mass with multiple areas of necrosis and hemorrhage.

**Fig. 2.** Hematoxylin and eosin stain shows a proliferation of atypic cells with hyperchromatic nuclei and eosinophic cytoplasm.

**Fig. 3a.** MRI: A high-intensity tumor on T2-weighted images, which was heterogeneously enhanced on gadolinium-enhanced imaging.

**Fig. 3b.** CT-SCAN: An irregular mass filling the pelvis with heterogeneous enhancement after PDCI.
Intensive chemotherapy and less extensive surgery could lead to favorable disease-free and overall survival rates. Clinical and radiologic assessment by digital rectal examination, cystoscopy with biopsies and CT scan, is generally carried after the first three to four cycles. However, a recent study by the soft tissue Sarcoma committee of the children’s Oncology Group (COG) proved that initial response is of no predictive value in group III (IRS). In patients with local disease, 5-year failure free survival (FFS) was 75% with frequent local relapse.

Tumor size >5 cm and stage T2 or more were independently predictive factors of worse FFS, while, non-embryonal histologies and large tumours tended to be associated with metastatic disease.

**Conclusion**

Rhabdomyosarcoma among young adult is a special and rare entity. A combination of more effective chemotherapy, earlier radiation therapy, and more bladder-sparing operations will be needed to improve the disease-free survival and bladder preservation rates of future patients.

**Authors’ contributions**

AS wrote and submitted the manuscript. TH, ZM and AM provided the images and MB reviewed the manuscript. WZ contributed to the writing and the reviewing of the manuscript. All authors read and approved the final manuscript.

**Patient consent**

Written consent was obtained from the patient to publish this case.

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**Declaration of competing interests**

The authors declare that there are no conflicts of interest regarding the publication of this article.

**References**

1. Lawrence Jr W, Anderson JR, Gehan EA, et al. Pretreatment TNM staging of childhood rhabdomyosarcoma: a report of the intergroup rhabdomyosarcoma study group. Children’s cancer study group. Pediatric Oncology group. Cancer. 1997;80:1165–1170.
2. Meza JL, Anderson J, Pappo AS, Meyer WH. Analysis of prognostic factors in patients with nonmetastatic rhabdomyosarcoma treated on intergroup rhabdomyosarcoma studies III and IV: the Children’s Oncology Group. J Clin Oncol. 2006;24(24):3844–3851.
3. Arndt C, Rodeberg D, Breitfeld PP, Raney RB, Ulrich F, Donaldson S. Does bladder preservation (as a surgical principle) lead to retaining bladder function in bladder/prostate rhabdomyosarcoma? Results from Intergroup Rhabdomyosarcoma Study IV. J Urol. 2004;171(6):2396–2403.
4. Assessment of response to induction therapy and its influence on 5-year failure-free survival (FFS) in Group III rhabdomyosarcoma (RMS): intergroup Rhabdomyosarcoma Study (IRS)-IV experience. J Clin Oncol. 2007 Nov 1;25(31):4909–4912.
5. Rodeberg DA, Anderson JR, Arndt CA, et al. Comparison of outcomes based on treatment algorithms for rhabdomyosarcoma of the bladder/prostate: combined results from the children’s Oncology group, German cooperative soft tissue sarcoma study, Italian cooperative group, and international society of pediatric Oncology malignant mesenchymal tumours committee: international results for treatment of BP RMS. Int J Canc. 2011;128(5):1232–1239.