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Adult bladder rhabdomyosarcoma: A case report

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ABSTRACT

Rhabdomyosarcomas (RMS) are mesenchymal tumors that develop at the expense of striated muscle fibers. It accounts for 6% of childhood malignancies. Rhabdomyosarcomas of the genitourinary tract also occur in children but are distinctly uncommon in adults. We report a case of bladder rhabdomyosarcoma in a 72-year-old patient who presented with gross hematuria and discuss difficulties of diagnosis and treatment.

1. Introduction

Rhabdomyosarcomas (RMS) are mesenchymal tumors that develop at the expense of striated muscle fibers.[1] It accounts for 6% of childhood malignancies and is, by a wide margin, the most common sarcoma of childhood.[2] The genito-urinary localization is seen in 20% of the cases of infantile rhabdomyosarcoma.[2] Genitourinary tract rhabdomyosarcoma in adults is very rare with a few sporadic cases reported in the literature. We report a case of bladder rhabdomyosarcoma in a 72-year-old patient who presented with gross hematuria and discuss difficulties of diagnosis and treatment.

2. Case report

A 72-year-old patient, diabetic, hypertensive, smoker, who consulted for terminal hematuria evolving for one year. There was no notion of professional exposure to aromatic amines or pelvic irradiation. He was in good general condition and had no hypogastric mass or inguinal adenopathy. The rectal examination was unremarkable. The urine culture was negative, the hemoglobin level was 10.2g/dl, the creatinine level was 11.7mg/L. Ultrasound of the urinary tract showed a 9 cm × 7 cm bladder tissular formation with dilatation of the right pyelocaliceal cavities (Fig. 1). Cystoscopy showed a bulging tumor with a wide implantation base involving the trigone and the right wall infiltrating the right ureteral orifice. We performed a transurethral resection of the bladder (TURB). The histological analysis of the resection chips showed a malignant sarcomatous proliferation of rounded and sometimes spindle-shaped cells. There were rhabdomyoblasts characterized by moderate nuclear atypia with sparse and sometimes banded cytoplasm. Mitoses were rare; the stroma was sparse and vascular (Fig. 2). Immunohistochemistry was negative for Desmin and positive for myogenin. Thus, the diagnosis of intermediate-grade embryonal RMS of the bladder was retained. As part of the extension work-up, a thoraco-abdominopelvic CT scan showed a tumor of the posterior wall of the bladder with the invasion of the perivesical fat. There was a right ureterohydronephrosis and pulmonary metastasis. After a decision of multidisciplinary consultation meeting and discussion, the patient was referred to the medical oncology department, where he received carboplatin-based chemotherapy. After three months, the patient died with multi-visceral failure.

3. Discussion

Bladder sarcomas are rare, representing less than 2% of all urogenital malignancies.[1] Sarcomas are largely dominated by leiomyosarcomas (50%) and rhabdomyosarcomas (20%), other histological types such as osteosarcomas or neurosarcomas being exceptional.[1] In our patient, histological examination coupled with immunohistochemistry allowed the diagnosis of intermediate grade embryonal RMS of the bladder. Although rare, RMS, especially the embryonal subtype, are most often described in children and adolescents.[2] Indeed, SMIs represent 5% of childhood cancers; the bladder being the most frequent location. RMS are most often reported in males with a sex ratio of 2.[2] There are no specific clinical signs of bladder RMS. In children, the earliest clinical sign appears to be urinary retention, whereas, in adults, hematuria is the most common inaugural sign.[1,3] Dysuria, constipation and pelvic pain appear later. Bladder RMS is most often located on the bladder floor and trigone, in contrast to leiomyosarcoma, which usually develops on the dome.[3]

In pediatric patients, the prognosis for patients with bladder and

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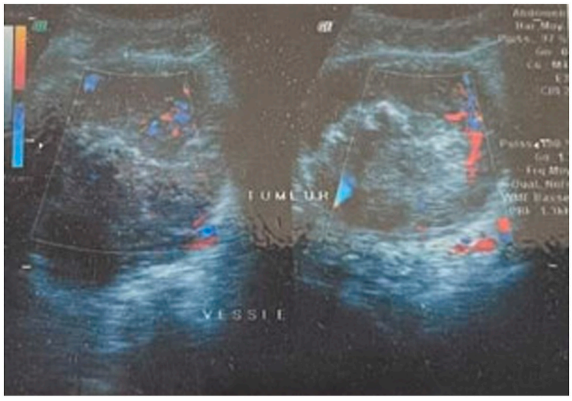

Fig. 1. Bladder ultrasound showing a tumor in the trigone and the right wall.

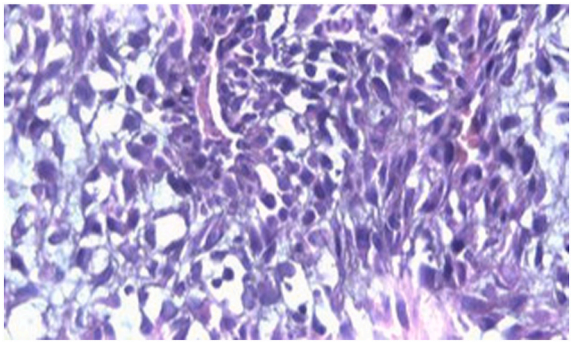

Fig. 2. Microscopic aspect of bladder Rhabdomyosarcoma (HE x 100).

prostate embryonal rhabdomyosarcoma is less favorable than for patients with disease in other sites, including the vagina, uterus, and paratesticular region (70%–73% and 84%–89%, respectively.[2] However, the prognosis and optimal management of adult embryonal rhabdomyosarcoma is uncertain owing to its rarity. There is no codified treatment allowing a better survival. Several treatments have been proposed but it turns out that a multimodal treatment seems necessary.

The therapeutic options include total or partial cystectomy, transurethral resection of the bladder, chemotherapy and radiotherapy.[2,4] In our case, RMS was diagnosed at the metastatic stage, which is why we opted for chemotherapy after TURB, which allowed the cessation of hematuria. In case of localized RMS, the gold standard remains radical cystectomy associated with bilateral lymph node dissection.[3] This radical surgery may be preceded by neo-adjuvant chemotherapy.[4] Radiation therapy adjuvant to surgery gives excellent results in case of embryonal RMS of the bladder in children.[5] Partial cystectomy can be proposed in young subjects who have a desire to father a child, when the tumor is small (less than 3 cm) and located at the level of the dome but it is imperative to ensure healthy surgical margins.[5] Regardless of the tumor stage at the time of diagnosis and regardless of the treatment, RMS are tumors with a poor prognosis with a survival of 3–19 months after the start of treatment.

4. Conclusion

Bladder rhabdomyosarcomas are rare mesenchymal malignancies in adults with a poor prognosis. Cystectomy represents the reference treatment for localized forms. The role of chemotherapy and radiotherapy in the management of metastatic forms remains to be evaluated.

Declaration of competing interest

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

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