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CASE REPORT

A case of prostate embryonal rhabdomyosarcoma in an adult patient

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Abstract

Prostate embryonal rhabdomyosarcoma (ERMS) is characterized by a high degree of malignancy, both local rapid growth with formation of large pelvic masses, often leading to renal failure due to urethral obstruction, and systemic spread, commonly to the lungs, liver, and bone. ERMS of the prostate is a commonly occurring tumor in infants and children. It is rarely seen in adults. Here, we report on a case of the prostate ERMS in a 27-year-old man.

Key words: prostate; embryonal rhabdomyosarcoma; surgery

Rhabdomyosarcoma (RMS) is a commonly occurring cancer in children, representing more than 50% of all soft tissue sarcomas. However, RMS of the prostate is a very rarely found malignancy in adults [1]. Histologically, three subtypes of RMS have been identified: alveolar histiotypic, pleomorphic, and embryonal growth pattern, including its rare spindle cell variant [2]. To date, there have been few reports on sarcoma manifesting as prostate ERMS. Clinical experience in treating adult RMS is limited to isolated cases reported in literature [3]. Here, we present a case of an adult patient with ERMS of the prostate.

Case report

A 27-year-old male presented with history of progressively increasing dysuria and obstructive voiding symptoms for the past 1 month. Fifteen days ago, he found blood in urine. Hemogram and biochemical parameters were within normal limits, without fever, back pain, nausea, vomiting, and other symptoms. BLD was 200 cells/μL. Prostate specific antigen (PSA) concentration was 1.10 ng/mL. Digital rectal examinations: Touched huge and hard mass, inactivities, boundary touch unclear. Ultrasound examination revealed irregular hypoechoic mass in the prostate area. A pelvic computed tomography (CT) scan was then performed, disclosing a 5.8 cm × 5.0 cm × 4.5 cm heterogeneous soft tissues mass rising from the prostrate, with disruption of the prostate anatomy and suspected infiltration of the bladder base and rectal wall. The patient underwent a transrectal biopsy of the prostate before he underwent suprapubic prostatectomy.

Pathological findings

Transrectal biopsy of the prostate revealed spindle cell malignant tumors, indicating spindle cell rhabdomyosarcoma. Immunohistochemical staining was positive for Vim, SMA, Des, h-caldesmon, Myogenin, CD99, bcl-2, and CD45, but negative for EMA, CK, S100, and CD34. Postoperative pathology showed prostate ERMS. The tumor was composed of small round cells and spindle shaped cells and rhabdomyoblasts were visible. Immunohistochemical staining was positive for Myogenin, Vim, CD99, Des, and WT-1, but negative for MyoD1 and CK.

Therapy and further course

A midline, extraperitoneal, lower abdominal incision was made extending from the umbilicus to the pubis. A huge, solid, and hard tumor, with rough surface, occupying most of pelvic, infiltrated bladder neck and bilateral seminal vesicle without infiltrating the rectum. Surgical

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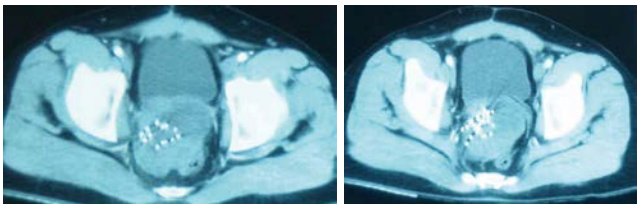

Fig. 1 CT showing radiotherapy (^{125}I seeds implantation)

options consisted of complete excision of the prostate tumor and bilateral seminal vesicle, partial resection of the bladder neck, and monitoring the protection of the rectum. The patient underwent chemotherapy (Vincristine, Actinomycin D, Cyclophosphamide) after surgery. However, after ten months, tumor recurrence was observed by clinical and CT examination. Tumor size was approximately 6.7 cm × 5.5 cm × 4.0 cm. Positron emission tomography-computed tomography (PET-CT) showed that the tumor was limited to the pelvic region without organ metastasis .Then the patient underwent radiotherapy (^{125}I seeds implantation); however, two months later, the tumor mass did not reduce. The patient reported with body pain, dysuria, and constipation. His general condition deteriorated and he had to take oral painkillers and laxative. This patient's condition was followed-up for 2 years.

Discussion

The incidence of prostate sarcoma is reported to be 0.1%–0.2% among primary prostate malignancies in adults; long-term survival for adult patients with prostate sarcoma is poor [4]. It grows rapidly and partially invades adjacent tissues, such as the bladder base or rectal wall. Patients with prostate rhabdomyosarcoma mostly present with dysuria or obstructive micturition problems [5]. In some patients, the compression of the rectum can cause constipation, rectal bleeding, and a sense of rectal fullness [6].

There is no pathognomonic radiological finding for prostate ERMS, and CT scan and MRI study revealed a large soft tissue mass with necrotic regions replacing the whole prostate; however, the radiological differential diagnosis with prostate adenocarcinoma can be very difficult [7]. The diagnosis is usually performed with a transrectal, perineal, or transurethral biopsy. Our patient underwent transrectal biopsy, and the histopathological analysis revealed spindle cell RMS of the prostate. Despite surgical contraindications, he underwent suprapubic prostatectomy. Postoperative pathology showed prostate ERMS. N J Sebire [8] reported that the immunohistochemical staining of pediatric rhabdomyosarcomas, with antibodies against MyoD and myogenin, provides sensitive and specific diagnostic information. Myogenin immunostaining is usually more clinically useful because it is more consistent and is associated with less nonspecific staining. Besides, younger age, normal PSA, large tumor volume, and poorly differentiated tumors help indicate prostate sarcoma.

The prognosis for patients with prostate ERMS is generally poor. In a clinical situation of an organ-confined disease, radical surgical extirpation should be performed [9]. The goal of surgery should be complete removal of disease, which usually requires radical cystoprostatectomy and pelvic extentaration [10]. As an alternative to surgery, radiotherapy and chemotherapy might be useful to achieve local treatment with organ function preservation and good quality of life. The major role of chemotherapy in this tumor location is the reduction of the primary tumor mass in order to assist with local therapy as well as to avoid tumor progression and the development of distant metastases [11]. Radiochemotherapy, alone as a local treatment, seems to be optimal especially in patients with smaller tumors and a good response to neoadjuvant chemotherapy [12]. But there are several relevant concerns about chemotherapy. Fryer reported that radiation therapy contributes to post-treatment morbidity. Another disadvantage is the fact that surgery is even more complicated after radiotherapy [13].

In conclusion, prostate ERMS requires a multidisciplinary treatment approach. At present, there is no standard treatment of prostate RMS in adults due to small number of cases. Some tumors in patients with postradiation sarcoma may be refractory to treatment with chemotherapy[1]. We, therefore, suggest that early radical resection prior to chemotherapy must be considered for such patients. Chemotherapy combined with radiation therapy should be adjunct to surgery to prevent local and distant recurrences. The severity of the disease needs to be informed to the patients and their family members before surgery and the patients need close follow-up after surgery.

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