Gazi Tıp Dergisi / Gazi Medical Journal

OLGU SUNUMU - CASE REPORT

2007: Cilt 18: Sayı 2: 86-88

REKTAL İNVAZYONLA SEYREDEN ERİŞKİN AGRESİF PROSTAT RABDOMİYOSARKOMU

İlker ŞEN[1], Metin ONARAN[1], Güldal YILMAZ[2], Leyla MEMİŞ[2], Lütfi TUNÇ[1], Hasan BİRİ[1], İbrahim BOZKIRLI[1]

Embryonal prostate sarcomas constitute less than 0.1% of primary prostate cancer cases in adults. Patients usually present with obstructive voiding and rectal compression symptoms. Due to the unusual aggressive behaviour of this disease, most patients presenting with advanced stage and metastatic lesions will have a poor prognosis. We report a patient with rectal perforation in which pelvic exenteration was performed in order to reach negative surgical margins. In addition, severe rectal bleeding leading to acute blood loss supported the indication to perform this radical surgery in our case. Thus, every physician should keep this rare disease in mind during the differential diagnosis of a patient sharing clinical features with our case to reach an exact diagnosis as soon as possible before metastatic disease develops.

Key Words: Prostate, embryonal rhabdomyosarcoma, tumor.

AN AGGRESSIVE ADULT PROSTATE RHABDOMYOSARCOMA WITH RECTAL INVASION

Embriyonal prostat sarkomları erişkin primer prostat kanserlerinin %0.1'den azını oluşturmaktadır. Hastalar genellikle obstrüktif idrar yapma ve rektal basılanma semptoları ile başvururlar. Agresif bir tümördür ve bir çok hasta prognozu kötüleştiren ileri evre ve metastatik lezyonlar ile başvurur. Çalışmamızda rektal perforasyon nedeniyle lokal kontrolü sağlamak için pelvik ekzantrasyon yapılan bir prostat embriyonal rabdomyosarkomu olgusu sunulmuştur. Ayrıca ciddi rektal kanamanın neden olduğu akut kan kaybı da radikal cerrahi seçiminde etkili olmuştur. Her klinisyen, nadir görülen ve mümkün olduğunca metastatik olmadan, doğru tanı konması gereken bu hastalıkla ilgili sunduğumuz klinik belirtileri ayırıcı tanıda akılda tutmalıdır.

Anahtar Kelimeler: Prostat, embriyonal rabdomiyosarkom, tümör.

Gazi Üniversitesi, Tıp Fakültesi, Üroloji[1] ve Patoloji[2] AD, Ankara, Türkiye

INTRODUCTION

Embryonal prostate sarcomas constitute less than 0.1% of primary prostate cancer cases in adults (1). Patients usually present with obstructive voiding and rectal compression symptoms. Due to the unusual aggressive behavior of this disease, most patients presenting with advanced stage and metastatic lesions will have a poor prognosis. Here we report embryonal rhabdomyosarcoma with rectal invasion in an adult.

CASE REPORT

A 30-year-old male presented with a history of progressive difficulty in voiding and defecation together with perineal pain for the previous 2 months. He was admitted to another hospital with the diagnosis of acute prostatitis and received antibiotic treatment with fluoroquinole for 14 days. When the symptoms did not improve, the patient was referred to our clinic. His general examination was unremarkable and hematological parameters were normal. Digital rectal examination revealed a very tender, smooth and large prostate with a PSA value of 1.5 ng/ml. Transrectal ultrasound of the prostate confirmed that the prostate volume was 220 cc with multiple hypoechoic lesions, and the patient underwent transrectal ultrasound guided biopsy of the prostate. Pathological examination revealed mononuclear cell infiltration with no tumoral cells but they reported that the biopsy material was insufficient to allow a conclusion. Contrast-enhanced abdominopelvic computerized tomography (CT) revealed a large and heterogeneous prostate containing central cystic-necrotic areas with lymphadenopathy in the right perivesical area (Figure 1a, 1b). Metastatic workup including thorax CT and bone scan detected no metastases in the liver, bone or lungs. The patient's complaints were progressive and he underwent a transurethral biopsy of the prostate. The pathology was embryonal rhabdomyosarcoma of the prostate. In the clinical follow-up, the patient suffered profuse rectal bleeding that led to acute blood loss, and rectoscopy revealed rectal invasion of the disease. Because of severe blood loss and local invasiveness, the patient underwent pelvic exenteration with ileal conduit and terminal colostomy. Sperm cryopreservation was not performed before the procedure because the patient had two children and cryopreservation was illegal in this country.

On gross examination, the tumor showed central hemorrhage and necrosis as it extended towards the bladder and rectum. In the latter, the tumor was penetrating the rectal wall, bulging into the lumen, and a lymphadenopathy was observed in the presacral area (Figure 1c). Microscopically, the tumor displayed hypercellular foci alternating with hypocellular areas and areas of myxoid change (Figure 2a). The tumor cells were small with scanty cytoplasm and had round to oval hyperchromatic nuclei (Figure 2b). Immunohistochemical studies revealed that the tumor cells stained

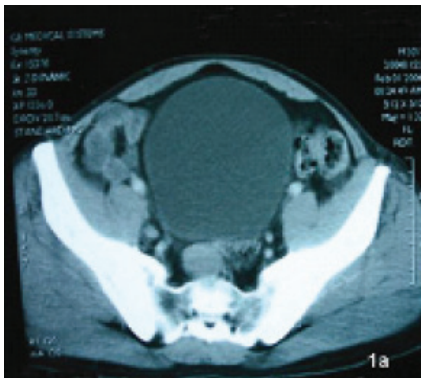

Figure 1a: Computerized tomography of the pelvis revealing a lymphadenopathy in the right perivesical space.

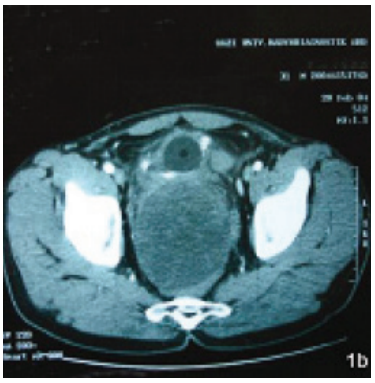

Figure 1b: Contrast-enhanced computerized tomography of the pelvis revealing a large and heterogeneous prostate containing central cystic-necrotic areas.

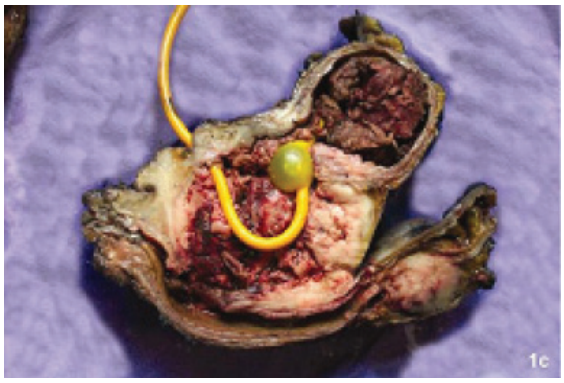

Figure 1c: Gross examination revealing the tumor with central hemorrhage and necrosis as it extended towards the bladder and penetrated the rectum, bulging into the lumen, and a lymphadenopathy in the presacral area.

positively with desmin and actin (Figure 2c). One month after surgery, the patient received adjuvant chemotherapy including adriamycine (60 mg/m2), ifosfamide (1.8 g/m2) and radiotherapy. After 14 months, he was reoperated for ileus and, because of ileal loop injury during this operation, bilateral permanent nephrostomy tubes were inserted. The patient has been living with no evidence of disease for 20 months.

DISCUSSION

Embryonal prostate sarcomas constitute less than 0.1% of primary prostate cancer cases in adults (1). Leiomyosarcomas are the most common subtype in adults, and rhabdomyosarcomas generally appear in pediatric patients with a median age of 5 years and display better prognosis (2, 3). This tumor is thought to arise from nonepithelial mesenchymal tissues of the prostate (4). Patients usually present with obstructive voiding and rectal compression symptoms, and the principal metastatic sites are the lungs, liver and skeleton, which are mainly osteoclastic (4).

The diagnosis of prostate sarcoma is difficult due to its wide age range, rapid progression, unusual presentation, normal PSA values and rarity. Lack of awareness of this entity in adults will eventually lead to delay in diagnosis, and the importance of diagnosis of this disease arises from a natural history of rapid progression because most patients presenting with advanced stage and metastatic lesions will have a poor prognosis (5). The tumor has a tendency to invade adjacent tissues like the ureters, leading to progressive uremia (6) or infiltration of bladder base and perirectal tissue planes penetrating the rectal lumen, like in our case.

Diagnosis of prostate sarcoma is usually achieved by transrectal biopsy of the prostate or by transurethral resection performed for relieving obstructive voiding symptoms. Clinical imaging can be enhanced by CT, which may reveal a large mass with areas of necrosis and cysts within the prostate. On the other hand, magnetic resonance imaging (MRI) might further enhance the clinical situation, especially when radical surgery is planned by cross-sectional pelvic imaging. MRI seems to provide a better soft tissue delineation and evaluation of tumoral mass with surrounding pelvic viscera (7).

Diagnosis is confirmed by histopathologic evaluation of the specimen. Microscopically, the tumor displays immature cells with pleomorphic nuclei and scanty cytoplasm (4). Immunohistochemical studies show positive staining with vimentin, actin and desmin (8, 9).

There is no established consensus on the treatment of adult rhabdomyosarcoma of the prostate due to the rarity of this disease and its poor prognosis (10). Multimodality treatments

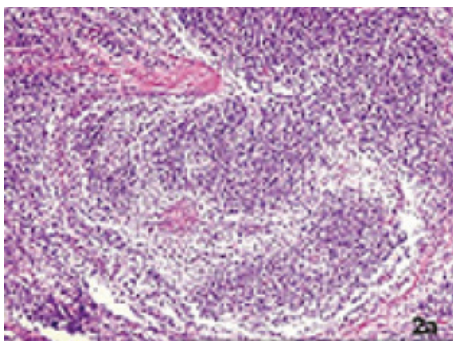

Figure 2a: Hypercellular foci of tumor alternating with hypocellular areas and myxoid change (HE X40).

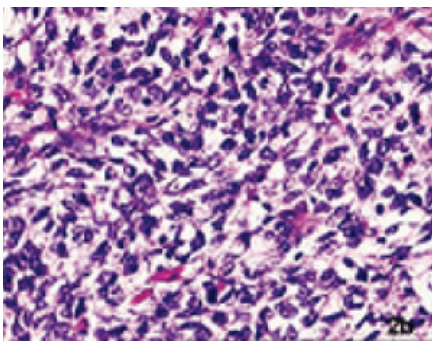

Figure 2b: Small tumor cells with scanty cytoplasm and round to oval hyperchromatic nuclei (HE X200).

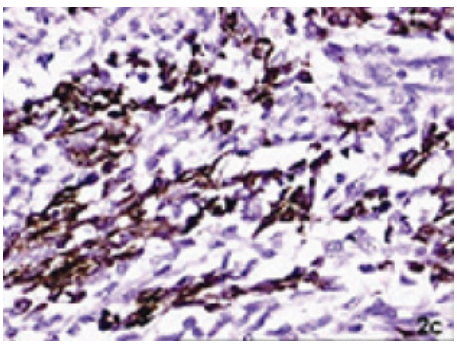

Figure 2c: Tumor cells show cytoplasmic positivity with desmin (Desmin X200).

including chemotherapy and radiotherapy adjunct to surgery should be applied. In addition, improved chemotherapeutic regimens together with more aggressive surgery might improve the prognosis of patients just like in children. Sexton et al. stated that negative surgical margins and absence of metastatic disease at presentation were predictive of long-term survival but tumor size, grade and histological subtype of the prostate sarcoma did not influence the survival outcome in these patients (10). The patient in the current study underwent pelvic exenteration because of rectal perforation in order to reach negative surgical margins. In addition, severe rectal bleeding leading to acute blood loss supported the indication to perform this radical surgery in our case. Thus, every physician should keep this rare disease in mind during the differential diagnosis of a patient sharing clinical features with our case to reach an exact diagnosis as soon as possible before metastatic disease develops.

Correspondence Address
Dr. İlker ŞEN
Yeniziraat mahallesi13. sokak 12/12
Dışkapı / Ankara
e-mail: ilkersen@gazi.edu.tr, ilkersenmd@hotmail.com
Tel: +90 532 742 74 23, Fax: +90 312 223 05 28

REFERENCES

1. Tetu B, Srigley JR, Bostwick DG. Soft tissue tumors. In Bostwick DG: Pathology of the Prostate, Churchill-Livingston, New York, 1990, P:117-120.
2. Morse MJ, Whitmore WF. Genitourinary sarcomas in adults. In Shiu MH and Brennan MF: Surgical Management of Soft Tissue Sarcomas, Lea and Febiger, Philadelphia, 1989, P:189-207.
3. Lemmon WT Jr, Holland JM, Ketchman AS. Rhabdomyosarcoma of the prostate. Surgery 1966; 59: 736-740.
4. Waring PM, Newland RC. Prostatic embryonal rhabdomyosarcoma in adults: a clinicopathologic review. Cancer 1992; 69: 755-762.
5. Nabi G, Dinda AK, Dogra PN. Primary embryonal rhabdomyosarcoma of the prostate in adults: Diagnosis and management. Int Urol Nephrol 2002; 34: 531-534.
6. King DG and Finney RP. Embryonal rhabdomyosarcoma of the prostate. J Urol 1977; 117: 88-90.
7. Hanna SL and Fletcher BD. MR imaging of malignant soft-tissue tumors. Magn Reson Imaging Clin North Am 1995; 3: 629-632.
8. De Jong ASH, Van Kessel-Van Vark M, Albus-Lutter CE. Pleomorphic rhabdomyosarcoma in adults; immunohistochemistry as a tool of its diagnosis. Human Pathol 1987; 18: 298-303.
9. Molenaar WM, Oosterhius AM, Ramaekers FCS. The rarity of rhabdomyosarcoma in adults: a morphological and immuno-histochemical study. Path Res Pract 1985; 180: 400-405.
10. Sexton WJ, Lance RE, Reyes AO et al. Adult prostate sarcoma: The M.D. Anderson Cancer Center Experience. J Urol 2001; 166: 521-525.