Case Report

Pure small cell carcinoma of the prostate: case report

Prostatın pür küçük hücreli karsinomu: olgu sunumu

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ABSTRACT

Extrapulmonary small cell carcinoma arising in the prostate gland has been described in several case series and case reports. However, pure small cell carcinoma of the prostate is rare, and there are only a few reports in the literature describing the clinical features and management of this neoplasm. These tumors are highly aggressive and commonly manifest with visceral metastasis at the time of diagnosis. This report describes the diagnosis and management of a 73-year-old patient with small cell carcinoma of the prostate in light of the reviewed literature.

Key words: Carcinoma; prostate; small cell

ÖZET

Bir çok olgu serisi ve olgu sunumlarında prostat bezinden gelişen ekstrapulmoner küçük hücreli karsinom tanımlanmıştır. Fakat, prostatın pür küçük hücreli karsinomu nadirdir ve literatürde bu neoplazmın klinik özellikleri ve tedavisi ile ilgili çok az sayıda yayını bulunmaktadır. Bu türlerin bir hayli agresifir ve tanı esnasında genellikle iç organ metastazı mevcuttur. Burada, 73 yaşında küçük hücreli prostat kanseri olan bir olguda ilgili literatürün ışığı altında tanı ve tedavi yöntemleri sunulmuştur.

Anahtar sözcükler: Karsinom; küçük hücreli; prostat

Introduction

Small cell carcinoma of the prostate (SCPCa), also termed neuroendocrine SCPCa, was first described by Wenk et al.⁶ in 1977. It has been recognized as a rare histologic variant, occurring in only 0.5% to 2% of prostatic primary tumors. SCPCa is one of the more common sites of extrapulmonary small cell carcinoma (EP-SCC). The clinical features of SCPCa differ from those of adenocarcinoma of the prostate in that SCC has a predilection to produce visceral metastases, lytic bony lesions, and low amounts of serum prostate-specific antigen (PSA).⁴ These tumors are highly aggressive, with a median survival of 9-10 months and a 5-year survival of less than 1%.⁵ We review the available literature to gain additional insight into the diagnosis, treatment, and prognosis of this disease.

Case report

A 73-year-old man complaining of obstructive urinary symptoms was admitted to our clinic. He had no prominent family or medical history, except for hypertension and atherosclerotic heart disease. The serum prostate-specific antigen (PSA) level was 1.1 ng/mL, and a comprehensive metabolic panel and complete blood count were within normal limits. A digital rectal examination revealed a mildly enlarged prostate, which was suggestive of benign pathology. The patient underwent transurethral resection of the prostate after physical examination, routine laboratory testing and uroflowmetry and IPSS (score: 21) evaluation, which revealed complete bladder outlet obstruction. Histopathology of the prostate specimen revealed pure small cell carcinoma of the prostate (Figure 1). The immunostain was positive for neuron-specific enolase (Figure 2). The findings for chromogranin, cytokeratins, CD-99 and LCA were negative.

The patient was then sent to the Department of Radiation Therapy and Oncology. After staging with PET CT and MRI, which revealed...
normal findings, the patient was diagnosed with localized disease, and systemic chemotherapy with carboplatin and etoposide for 4-6 cycles was planned, followed by radiotherapy to the prostate. A chemotherapy regimen was started in May 2011 consisting of 6 cycles of carboplatinum (290 mg/day, once in 21 days according to the AUC 4 protocol) and etoposide (100 mg/m², 170 mg for 3 days in 21-day cycles), and the treatment was completed in September. The patient was otherwise healthy and stable, except for lung edema triggered by cardiac insufficiency, and treated in the same manner during this period. Currently, he is planned to undergo radiotherapy, after confirmation with a control PET CT that revealed normal findings.

Discussion

Primary small cell carcinoma of the prostate is uncommon and is usually discovered incidentally in concordance with histologic samples of adenocarcinomas. In approximately 50% of the patients, pathological findings reveal adenocarcinomatous elements.[4] Pure small cell carcinoma of the prostate is an extremely rare occurrence, with clinical features unlike those of prostatic adenocarcinoma.

The most typical age at diagnosis is 61 to 70 years, but some studies suggest a range from 24 to 90 years.[5] Our patient was 73 years old. Unlike typical prostate adenocarcinoma, which is asymptomatic in most cases, obstructive urinary tract signs are the most common symptoms, suggesting rapid growth with extracapsular extension. Our patient presented with urinary outflow obstruction. Other common symptoms include abdominal or pelvic pain and hematuria. The signs and symptoms of advanced/metastatic disease include hepatomegaly, lymphadenopathy (including pelvic masses), jaundice, ascites, and cachexia in end-stage disease.[5] Prostatic malignant neuroendocrine cells tend to produce ectopic peptides, with adrenocorticotropic hormone (ACTH) and calcitonin being detected most frequently in the serum. Approximately 10% of small cell carcinoma cases present with paraneoplastic syndromes.[5,6] Paraneoplastic syndromes are of a neurologic nature (confusion, sensory, or motor deficits) and can include peripheral neuropathy, Lambert-Eaton syndrome (myasthenia gravis), Cushing disease, and limbic encephalitis.[6] None of these symptoms were found in our patient. On digital examination, the prostate is unlikely to be distinguishable from typical prostatic adenocarcinoma. The prostate may be normal, as in our case, have a discrete nodule, or be irregular with multiple nodules (extra-
such as complications of brain and bone metastases. Regarding metastases, its role is palliative, as it may control local symptoms, also play a role in the treatment of patients with SCPCa and control and a potential survival benefit. Radiation therapy could with chemotherapy should be considered as a treatment strategy in local or early SCPCa. This modality may provide better local control and a potential survival benefit. Radiation therapy could also play a role in the treatment of patients with SCPCa and metastases. Its role is palliative, as it may control local symptoms, such as complications of brain and bone metastases. Regarding irradiation, no exact guidelines for the total dose or irradiation volume have been suggested. Because of the disease propensity for local and pelvic lymph node relapse, the treatment volume should include the pelvic lymph nodes, with a dose between 45 and 55 Gy (daily dose 1.8 Gy), followed by a boost to the prostate volume, which in some studies may reach a total dose of 72 Gy. The paradigm for the multimodal treatment of SCPCa is derived from the treatment for lung SCC and is consistent with cisplatin-based chemotherapy and radiation therapy. Various regimens adding cisplatin as the main agent have been used, including etoposide, cyclophosphamide, doxorubicin and vincristine; carboplatin and etoposide; carboplatin and taxanes; and cisplatin, ifosfamide, and etoposide. The role of hormonal treatment in SCPCa is controversial. In the setting of mixed histologies, hormonal therapy should be used according to stage. Hormonal therapy should not be used as the sole therapy in SCPCa. Extensive bone metastases can be treated with either samarium-153 or radiolabeled indium-111-somatostatin analogs. Rapid and significant pain relief can be achieved. After octreoscan positivity for SCPCa, somatostatin analogs and their radionuclide derivatives might exert a palliative effect in symptomatic patients.

The prognosis of SCPCa is poor, even in localized and early cases, and the median survival is limited to months. Long-term survivors are rare. The best survival chances seem to be achieved with concurrent external radiation therapy and cisplatin and etoposide. Because of the grim prognosis, innovative methods targeting basic molecules and the induction of the host immune response should be developed.

Conflict of interest
No conflict of interest was declared by the authors.

References