Kas tutulumu olmayan dev sarkomatoid mesane karsinomlu bir hastada mesane koruyucu yaklaşım

Bladder sparing approach in a patient with giant non-muscle invasive sarcomatoid urothelial carcinoma of the bladder

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Abstract

Carcinosarcoma or sarcomatoid carcinoma of the urinary bladder has a poor prognosis with a short time from diagnosis to death despite aggressive treatment. In this case, we report a 73 years-old man with the pathological diagnosis of sarcomatoid urothelial carcinoma of urinary bladder who survived without recurrence for 27 months after bladder sparing treatment and died due to cerebrovascular disease.

Key Words: Sarcomatoid, urinary bladder, urothelial carcinoma

Özet

Mesanenin karsinosarkom veya sarkomatoid karsinomu kötü prognozlu olup, agresif cerrahi tedaviye rağmen hastalar kısa sürede kaybedilmektedir. Bu makalede patolojisi sarkomatoid ürotelyal karsinom gelen 73 yaşında erkek hastada, mesane koruyucu yaklaşımla yapılan tedavi ve hastalıksız olarak geçen 27 aylık süreç sonrası beyin damar hastalığı nedeniyle ölen olgu sunulmuştur.

Anahtar Kelimeler: Mesane, sarkamatoid, ürotelyal karsinom

Introduction

Carcinosarcomas are unusual, primary, malignant, nonurothelial tumors of urinary bladder, histologically biphasic, characterized by an intimate admixture of malignant epithelial elements (carcinoma) and malignant mesenchymal elements (sarcoma). The patients with carcinosarcoma or sarcomatoid carcinoma of urinary bladder have poor prognosis despite aggressive treatment. In this case, we report a patient with sarcomatoid urothelial carcinosarcoma who was followed-up without tumor recurrence for 27 months after bladder sparing treatment approach despite of large volume of tumor without muscular invasion.

Case report

A 73 years-old man who had hematuria for two weeks was referred to our hospital. The ultrasonographic evaluation of the patient revealed a 75 mm polipoid mass. Computerized tomography (CT) showed a 74x65x60 mm mass with wide base of 33x30mm occupying the anterior of left ureteral orifice and projecting to the lumen with a 13mm thick pedicle and had a soft tissue density in contrast medium (Figure 1). There was no sign of local invasion, lymph node or distant metastasis in CT images of torax and abdomen. Also, there was no bone metastasis detected in sintigraphic evaluation. Cystoscopic examination revealed a solid tumor occupying the left wall of the bladder. Transurethral resection was performed and 160gr specimen was resected in 2 hours. Bimanuel examination revealed no palpabl mass. Intravesical 40mg mitomycin C was installated in postoperative 24 hours.

Predominantly fusiform, sarcomatoid tumor cells with eosinophilic cytoplasm were detected in the histolo-

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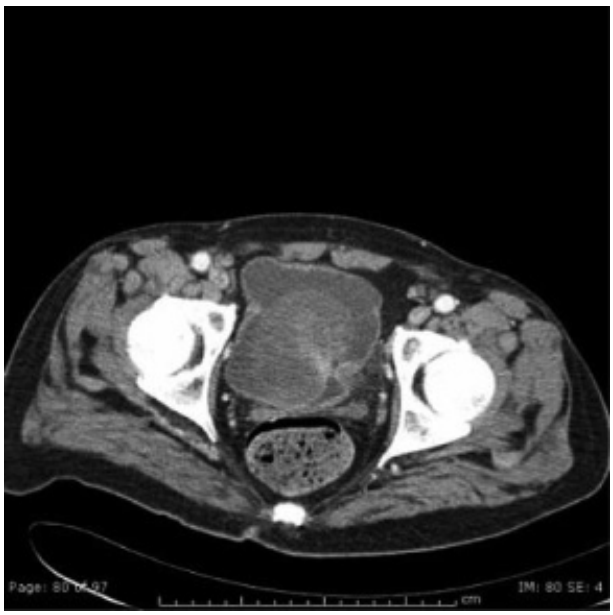

Figure 1. The CT images of the tumor and pedicle

gical slides obtained from samples of tumor material (Figure 2). Less amount of epithelloid tumor cells with oval-round nucleolus and large cytoplasm were also present. In the immunohistochemical examination cytokeratin staining was positive for those epithelloid cells and particularly some fusiform cells (Figure 3). In "tumor base", low grade papillar urothelial carcinoma, focal dysplasia and carcinoma in situ were detected in a small area. Vimentin applied by immunohistochemical method was positive in tumor cells (Figure 4), whereas Desmin, S100, smooth muscle actin (SMA) were negative. There was lamina propia invasion in these samples with no muscular invasion. This case was considered as sarcomatoid urothelial carcinoma according to histopathological and immunohistochemical results.

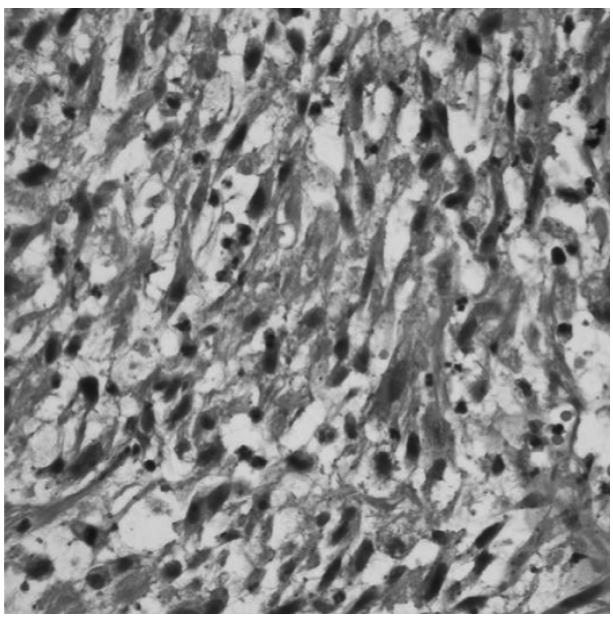

Figure 2. Sarcomatoid tumor cells with fusiform,eosinophilic cytoplasm (HE×400)

The patient underwent repeat transurethral resection after 4 weeks from the first resection. Pathological examination showed no tumor in the resected specimen .

Intravesical Bacillus Calmette-Guerin (BCG) was installed for 6 weeks with a dose of 81mg per week. Then the patient was followed-up for 2 years with cystoscopic evaluation in 3 months period and CT once a year. The maintenance BCG was not installed for the patient. No recurrence was detected. Patient died due to cerebrovascular disease 27 months after the diagnosis.

Discussion

Carcinosarcoma of the bladder is a rare entity; approximately 80 cases have been reported. A few cases have been reported in the renal pelvis.[1] Epidemiologically, there is a male predominance with a male-to-female ratio of 4:1 and a peak incidence during the seventh decade of the life.[2] Owing to their abnormal effects on cell replication, pelvic radiotherapy and cyclophosphamide therapy might be possible etiologies of carcinosarcomas.[2,3,4] Symptoms at presentation do not differ from those of other bladder cancers; hematuria is the main symptom.[2] Our patient was a man in his seventh decade of life, and hematuria was the only symptom. He had no history of pelvic radiotherapy or cyclophosphamide therapy. Although no consensus on the histogenesis and nomenclature exists, the terms carcinosarcoma or sarcomatoid carcinoma have been used. Pathologically, carcinosarcoma originates from the mesenchymal and epithelial components of the bladder, and tumors are an admixture of malignant epithelial and mesenchymal components. Although the carcinomatous component is positive for epithelial markers like epithelial membrane antigen and cytokeratin, the sarcomatous component is negative. The most common sarcomatous elements in carcinosarcoma are chondrosarcoma, leiomyosarcoma, and malignant fibrous histiocytoma, followed by osteosarcoma, fibrosarco-

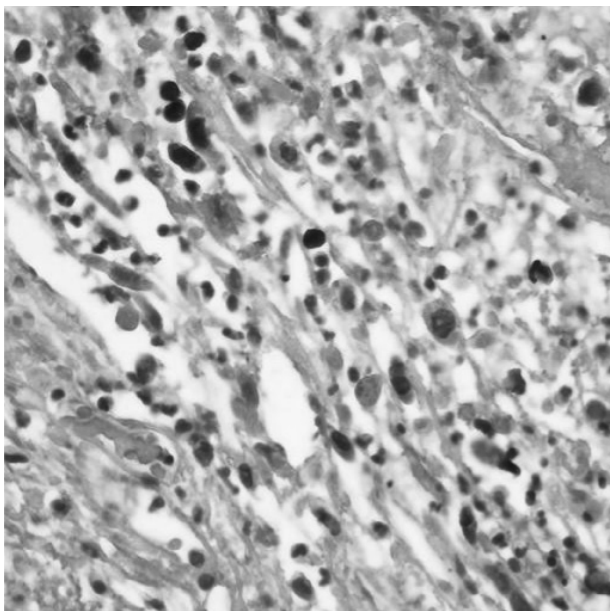

Figure 3. Keratin positivity in sarcomatoid areas (Keratin×400)

ma, and rhabdomyosarcoma.[5] In the tumor presented by this case, epithelloid or fusiform cells showed positive reaction by keratin and vimentin together with the presence of typical urothelial carcinoma areas. Thus the tumor was considered as sarcomatoid carcinoma.

Genetic analysis shows chromosomal abnormalities like loss of heterozygosity on the short arm of chromosome 9 (9p21), which is the chromosomal localization

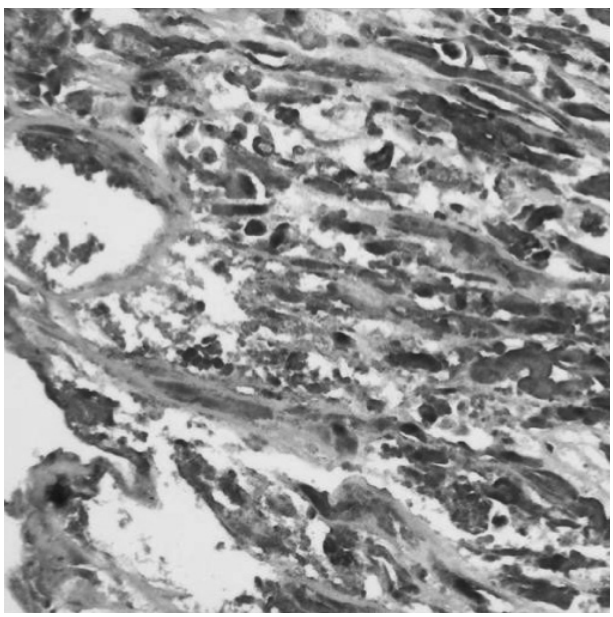

Figure 4. Vimentin positivity in sarcomatoid areas (Vimentin×400)

of the tumor-suppressor gene *TP*16 and on the long arm of chromosome 11.[4,6] The Mayo Clinic reviewed their more-than-50-year experience with 15 patients with carcinosarcoma and 26 patients with sarcomatoid tumors.[3] Both tumor entities shared a similar presentation in predominantly elderly men. The most common epithelial component in both was urothelial. Local extent correlated with outcome, yet most patients had locally advanced tumors at the time of diagnosis. Adversely, our patient had localized tumor at the time of diagnosis without any local extension. Radical cystectomy followed by adjuvant radiotherapy and/or chemotherapy is recommended as a treatment modality.[2,5] We did not perform radical surgery, radiotherapy or chemotherapy to our patient since the tumor had no invasion of the muscle layer. Only, intravesical BCG was performed to our patient for 6 weeks period because the pathological examination revealed carcinoma in situ. The outcome was poor in patients with carcinosarcoma and sarcomatoid tumors, with patients succumbing to their disease within 1 to 2 years despite aggressive surgical management. Patients affected by carcinosarcoma have a low survival rate, averaging 17.2 months.[3] Our patient lived for 27 months without tumor recurrence or metastasis and died due to another reason than the tumor.

Carcinosarcoma and sarcomatoid tumor is a rare but rapidly progressive disease of urinary bladder with poor prognosis. Immidiate and intensive treatment should be recommended after diagnosis. However, bladder preventive modalities might be an alternative for treatment in patients who have localized tumor without muscle invasion and unsuitable general status for radical surgery.

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