CASE REPORT

Biphasic and monophasic sarcomatoid carcinoma of the urinary bladder

Yoh Matsuoka, MD,[1,2] Makoto Hirokawa, MD,[1] Kimio Chiba, MD,[1] Takafumi Hashiba, MD,[1] Takeshi Tomoda, MD,[1] Shimpei Sugiura, MD[1]

[1]Department of Urology, Fujisawa City Hospital, Kanagawa, Japan

[2]Department of Urology, Graduate School, Tokyo Medical and Dental University, Tokyo, Japan

MATSUOKA Y, HIROKAWA M, CHIBA K, HASHIBA T, TOMODA T, SUGIURA S. Biphasic and monophasic sarcomatoid carcinoma of the urinary bladder. The Canadian Journal of Urology. 2008.15(3):4106-4108.

We report a rare case of sarcomatoid carcinoma of the urinary bladder which showed both of biphasic and monophasic phenotypes through its clinical course. A 64 year-old man presented with gross hematuria and interrupted voiding. Radiological examination demonstrated a pedunculated tumor in the bladder with no distant metastases. Pathological diagnosis of transurethral resection revealed biphasic sarcomatoid carcinoma, pT1, composed of squamous cell carcinoma component and spindle cell component. The disease had repeated local recurrences and partial and total cystectomies were performed consequently. The recurrent tumors were monophasic sarcomatoid carcinoma purely composed of spindle cell component. The patient died of local and metastatic disease 13 months after the diagnosis. We consider that sarcomatoid carcinoma should be resected radically at the initial surgical treatment because of aggressive potential of spindle cell component. To our knowledge, this is the first case in which biphasic sarcomatoid carcinoma of the bladder recurred as monophasic tumor. The pathological study and management of this neoplasm are discussed, and the literature is reviewed.

Key Words: immunohistochemistry, sarcomatoid carcinoma, spindle cell, urinary bladder

Introduction

Sarcomatoid carcinoma of the urinary bladder is an uncommon neoplasm accounting for 0.31% of all bladder malignancies, which has acquired sarcomatous phenotype arising from the epithelium.[1-3] Although the age distribution, male preponderance, and clinical symptoms are not distinctive from conventional urothelial carcinoma, sarcomatoid carcinoma behaves as a high-grade malignancy with extremely poor outcome. The survival rate has to be expressed in months rather than in years.[3] Sarcomatoid carcinoma can be classified into two categories; biphasic and monophasic, depending upon the presence or absence of a discernible carcinomatous component.[4,5] Herein, we report a rare course of biphasic sarcomatoid carcinoma of the urinary bladder, which recurred rapidly as monophasic type after each surgery.

Case report

A 64-year-old man presented with gross hematuria and interrupted voiding in April 1998. Computed tomography (CT) scan demonstrated a 6 cm x 6 cm pedunculated tumor with a peduncle 0.6 cm in diameter on the posterior wall of the urinary bladder, Figure 1. Distant

Accepted for publication April 2008

Acknowledgment
The authors thank Hitoshi Masuda and Gaku Arai from Department of Urology, Tokyo Medical and Dental University, for their editorial assistance.

Address for correspondence to Dr. Yoh Matsuoka, Department of Urology, Graduate School, Tokyo Medical and Dental University, 1-5-45 Yushima, Bunkyo-ku, Tokyo 113-8519, Japan

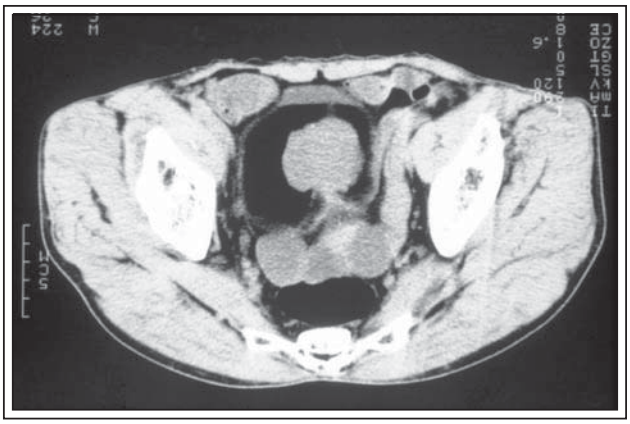

Figure 1. Computed tomography shows a pedunculated mass on the posterior wall of the urinary bladder.

metastases were not identified. Cystoscopic examination revealed a large polypoid tumor which blocked the internal urethral orifice. Transurethral resection (TUR) was performed and the weight of the resected tumor tissue was 85 g. Randomized bladder biopsy was not enforced because other areas were unremarkable and preoperative urine cytologies showed no abnormality. The histopathological diagnosis was sarcomatoid carcinoma variant of squamous cell carcinoma (SCC), pT1. A repeated cystoscopy 3 weeks after the TUR revealed a new polypoid lesion near the original tumor site. Partial cystectomy was then performed and the tumor infiltrated to the superficial muscular layer (pT2a). After 3 months, pelvic CT scan demonstrated a bulky polypoid bladder tumor again and total cystectomy was performed immediately. The recurrent tumor was 13 cm x 9 cm x 6 cm in size and invaded the perivesical fatty tissue, Figure 2. The patient underwent two courses

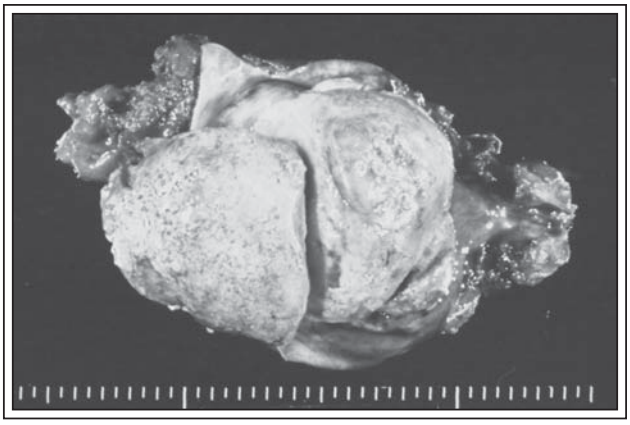

Figure 2. Gross specimen of total cystectomy weighed 452 g. A protuberant solid mass fills the urinary bladder completely.

of adjuvant chemotherapy consisting of methotrexate, epirubicin, and cis-platinum. Four months after the total cystectomy, CT scan identified metastases to pelvic lymph nodes and ischial bones. The metastatic lesions were resistant to radiation therapy and spread to the lung. The patient died 13 months after the diagnosis.

Pathologically, the tumor tissue obtained by the TUR was an admixture of high-grade SCC and mesenchymal-like elements. There were many mitotic cells in the latter which consisted of pleomorphic spindle shaped cells with variable amounts of cytoplasm and bizarre nuclei. The spindle cell component was focally arranged in a storiform pattern, partially immunostained for vimentin and keratin, Figure 3a, but negative for desmin, a smooth muscle actin, and S-100 protein. Tumor tissues obtained by the partial and total cystectomies were monophasic sarcomatoid carcinoma which consisted of only spindle cell component, immunostained positively for keratin and vimentin, Figure 3b. There was no conventional light-microscopic evidence of epithelial differentiation.

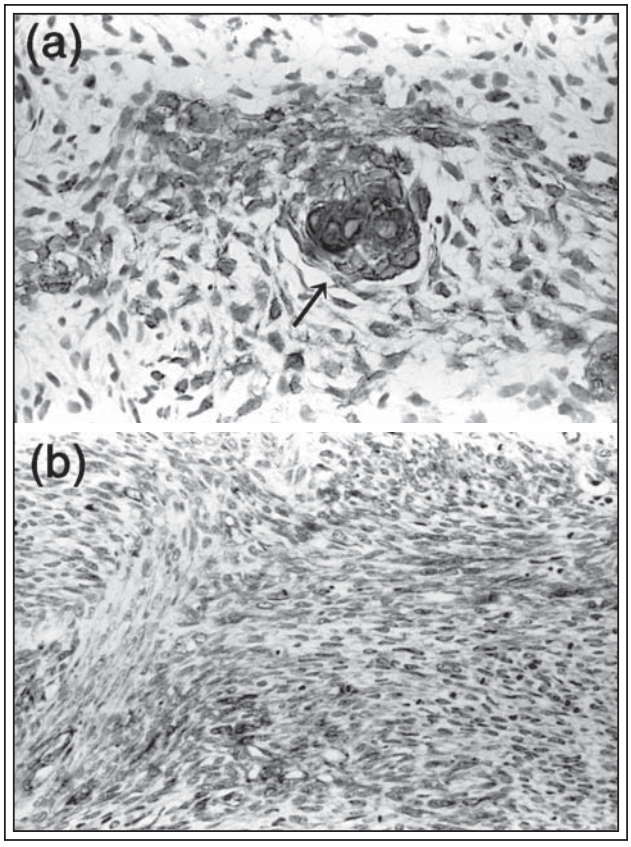

Figure 3a. Spindle shaped cells with foci of squamous cell carcinoma (arrow) appear positive with anti-keratin antiserum (original magnification X400). **3b.** Storiform arrangement of spindle cells showing immunoreactivity for vimentin (original magnification X200).

Comment

Sarcomatoid carcinoma is a malignant spindle cell variant of epithelial tumor, with or without coexistent overt carcinoma.[6] Although there is controversy as to whether carcinosarcoma and sarcomatoid carcinoma should be regarded as separate entities or not,[7] definite diagnosis can be obtained on the basis of immunohistochemical and electron microscopic findings that sarcomatous elements of sarcomatoid carcinoma have epithelial nature.[3] Recent genetic and molecular-based studies support the concept of a monoclonal origin of both carcinomatous and sarcomatous elements of sarcomatoid carcinoma and suggest that genetic divergence may occur during tumor progression and differentiation.[8] In sarcomatoid carcinoma of the urinary bladder, epithelial components recognized on light microscopy show variable phenotypes, with areas of moderately or poorly differentiated transitional cell carcinoma in most cases. Less frequently, foci of SCC, adenocarcinoma, and undifferentiated small or large cell carcinoma are seen.[3,6,9]

Among 63 patients with sarcomatoid carcinoma of the bladder in literatures, 54 patients (86%) were biphasic tumors in which both of apparent epithelial and mesenchymal components were identified by light microscopy; the rest (14%) were recognized as monophasic tumor composed of a pure sarcomatoid population of cells.[3,6,9,10] To our knowledge, no case has been reported in which biphasic sarcomatoid carcinoma recurred as monophasic tumor. In the present case, spindle cell component was existed in both of the initial tumor and recurrent tumors, while SCC component was identified only in the initial tumor. The tumor recurred as monophasic sarcomatoid carcinoma, suggesting that the proliferation of immature spindle cells was much more aggressive than that of SCC.

Sarcomatoid variants of bladder tumor tend to be bulky, protuberant, and polypoid with rapid growth, as the present case repeated polypoid growth.[9,11,12] Therefore, at first sight, the tumor seemed to be resectable by local excision such as TUR and partial cystectomy. This neoplasm, however, behaved much more aggressively, especially in recurrence, than conventional high-grade urothelial carcinoma with similar degree of invasion. Sejima et al[13] reported two cases of sarcomatoid carcinoma which had obvious local progression just after TUR. They died of local and metastatic disease a few months after the endoscopic surgery, whose clinical courses were similar to ours. Early local recurrence in these cases implies that spindle tumor cells have high potential of dissemination and implantation by surgical procedure.

Among the previously reported 63 patients, 23 cases (37%) were treated with TUR, 28 cases (44%) with total or partial cystectomy, 29 cases (46%) with radiation therapy, and 10 cases (16%) with chemotherapy.[3,6,9,10] Radiation therapy and chemotherapy were performed as adjuvant or palliative treatment. The prognosis was extremely poor regardless of treatment because most of the cases were in advanced stages at the initial visit, but a small number of cases have been reported with their prolonged survival by combined surgery and radiation or chemotherapy.[5,10,14] Lopez-Beltran et al[6] reported that mean survival of patients with this malignancy was 10 months, and pathological stage was the best predictor of the survival. We consider that extended radical surgery should be performed as the initial treatment for sarcomatoid carcinoma of the urinary bladder, even if the tumor is small, polypoid and/or in early stage. Repeat of conservative surgery should be avoided. □

References

1. Young RH. Carcinosarcoma of the urinary bladder. *Cancer* 1987;59:1333-1339.
2. Lahoti C, Schinella R, Rangwala AF, Lee M, Mizrachi H. Carcinosarcoma of urinary bladder: report of 5 cases with immunohistologic study. *Urology* 1994;43:389-393.
3. Torenbeek R, Blomjous CM, de Bruin PC, Newling DW, Meijer CM. Sarcomatoid carcinoma of the urinary bladder. *Am J Surg Pathol* 1994;18:241-249.
4. Nappi O, Glasner SD, Swanson PE, Wick MR. Biphasic and monophasic sarcomatoid carcinomas of the lung. *Am J Clin Pathol* 1994;102:331-340.
5. Wick MR, Ritter JH, Humphrey PA. Sarcomatoid carcinomas of the lung: a clinicopathologic review. *Am J Clin Pathol* 1997;108:40-53.
6. Lopez-Beltran A, Pacelli A, Rothenberg HJ et al. Carcinosarcoma and sarcomatoid carcinoma of the bladder: clinicopathological study of 41 cases. *J Urol* 1998;159:1497-1503.
7. Eble JN, Sauter G, Epstein JI, Sesterhenn I. Pathology and genetics of tumours of the urinary system and male genital organs. Lyon: IARC Press;2004.
8. Sung MT, Wang M, MacLennan GT et al. Histogenesis of sarcomatoid urothelial carcinoma of the urinary bladder: evidence for a common clonal origin with divergent differentiation. *J Pathol* 2007;211:420-430.
9. Young RH, Wick MR, Mills SE. Sarcomatoid carcinoma of the urinary bladder. *Am J Clin Pathol* 1988;90:653-661.
10. Serio G, Zampatti C, Ceppi M. Spindle and giant cell carcinoma of the urinary bladder: a clinicopathological light microscopic and immunohistochemical study. *Br J Urol* 1995;75:167-172.
11. Fromowitz FB, Bard RH, Koss LG. The epithelial origin of a malignant mesodermal mixed tumor of the bladder: report of a case with long-term survival. *J Urol* 1984;132:978-981.
12. Murata T, Soga T, Tajima K et al. Sarcomatoid carcinoma of the urinary tract. *Pathol Int* 1994;44:138-144.
13. Sejima T, Miyagawa I. Sarcomatoid carcinoma of the urinary bladder: 3 case reports. *Urol Int* 1999;62:51-54.
14. Froehner M, Gaertner HJ, Manseck A, Wirth MP. Durable complete remission of metastatic sarcomatoid carcinoma of the bladder with cisplatin and gemcitabine in an 80-year-old man. *Urology* 2001;58:799.