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Case Report

Multiple cytokeratin-negative malignant tumors composed only of rhabdoid cells in the renal pelvis: a sarcomatoid urothelial carcinoma?

Tadashi Terada

Department of Pathology, Shizuoka City Shimizu Hospital, Shizuoka, Japan

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Abstract: The author presents a unique case of multiple cytokeratin-negative malignant tumors consisting only of rhabdoid cells in the renal pelvis. A 54-year-old man complained of hematuria. A transurethral endoscopic examination revealed multiple papillary tumors, and transurethral resection of the bladder tumors was performed. Pathologically, they were ordinary papillary urothelial transitional cell carcinomas. Imaging modalities revealed multiple tumors of the right renal pelvis, and nephrectomy was performed. Grossly, three polypoid tumors measuring 2-4 cm were present in the pelvis. Histologically, they were composed only of malignant cells with rhabdoid features. There were no elements of transitional cell carcinoma. Immunohistochemically, the pelvic tumors were positive for vimentin and Ki-67 antigen (labeling=40%). They were negative for pancytokeratins (AE1/3, CAM5.2, KL-1 and polyclonal wide), 34βE12, cytokeratin (CK) 5/6, CK7, CK8, CK14, CK18, CK19, CK20, melanosome, EMA, CEA, desmin, S100 protein, α-smooth muscle actin, myoglobin, myogenin, CD34, p53 protein, p63, CD3, CD20, CD30, CD45, CD45RO, chromograin, synaptophysin, CD56, CD68, and KIT. NSE and PDGFRA were focally present, but this appeared nonspecific. Namely, the pelvic tumors expressed only vimentin. The author speculates that the pelvic multiple malignant "rhabdoid" tumors are not sarcomas but urothelial "rhabdoid" carcinoma with complete loss of CKs.

Keywords: Rhabdoid tumor, urothelial carcinoma, renal pelvis, immunohistochemistry

Introduction

It has been reported that urothelial carcinoma of renal pelvis infrequently shows rhabdoid features [1]. Several cases of sarcomatoid urothelial carcinoma have been reported in the literatures [2, 3]. Cytokeratin (CK) expression has been recognized in these cases [2, 3]. Malignant neoplasm of rhabdoid features have rarely reported in the renal pelvis [4-6]. Several types of sarcomas including rhabdomyosarcomas and lymphomas may occur in the renal pelvis [1, 7-9]. The author herein reports a unique case of multiple malignant "rhabdoid" tumors in the renal pelvis. The pelvic tumors in this case expressed only vimentin and are negative for CKs and other mesenchymal markers.

Case report

A 54-year-old man was admitted to our hospital because of macroscopic hematuria. A transurethral endoscopic examination revealed multiple papillary tumors in the urinary bladder, and transurethral resections of bladder tumors were performed. Pathologically, they were ordinary papillary urothelial transitional cell carcinomas without invasion (pTa). Later, imaging modalities including US, CT and MRI revealed multiple tumors of the right renal pelvis. Clinical cytology of right ureter urine showed atypical cells. Nephrectomy was performed.

Grossly, there were three polypoid tumors measuring 2-4 cm in the renal pelvis (**Figure 1**). Histologically, they were composed only of malignant cells with eosinophilic cytoplasm and eccentrically located nuclei (rhabdoid features) (**Figures 2**, **3** and **4**). Mitotic figures were scattered. No elements of urothelial transitional cell carcinoma were recognized in the tumor or in the urothelial layer near the pelvic tumors. No cytoplasmic filamentous inclusions were recognized. Nucleoli were present but not prominent. No invasive features were recognized.

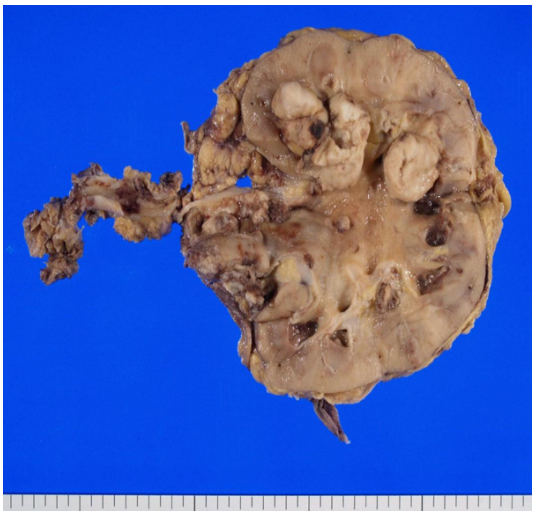

Figure 1. Gross findings of pelvic tumor. The renal pelvis contains multiple polypoid tumors.

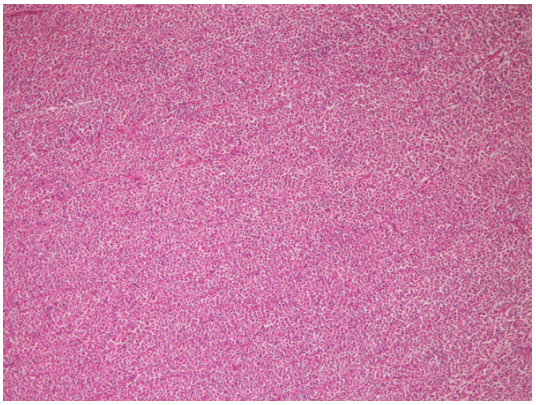

Figure 2. The carcinoma cells are medullary tumor without stroma. HE, x40.

An immunohistochimical study was performed, using Dako Envision method (Dako Corp., Glostrup, Denmark), as previously described [10, 11]. The antigens examined and results are shown in **Table 1**. The pelvic tumors were positive for vimentin (**Figure 5**) and Ki-67 antigen (labeling=40%) (**Figure 6**). They were negative for pancytokeratins (AE1/3, CAM5.2, KL-1, and polyclonal wide), 34βE12E12, CK5/6, CK7, CK8, CK14, CK18, CK19, CK20, melanosome, epithelial membrane antigen, carcinoembryonic antigen, desmin, S100 protein, α-smooth muscle actin, myoglobin, myogenin, CD34, p53 protein, p63, CD3, CD20, CD30, CD45, CD45RO, chromogranin, synaptophysin, CD56, CD68, and KIT. Neuron-specific enolase (**Figure 7**) and platelet-derived growth factor-α (**Figure 8**) were focally present. A diagnosis of multiple malignant "rhabdoid" tumors of the renal pelvis was made by the author. The patient is now alive without metastasis 9 months after the nephrectomy.

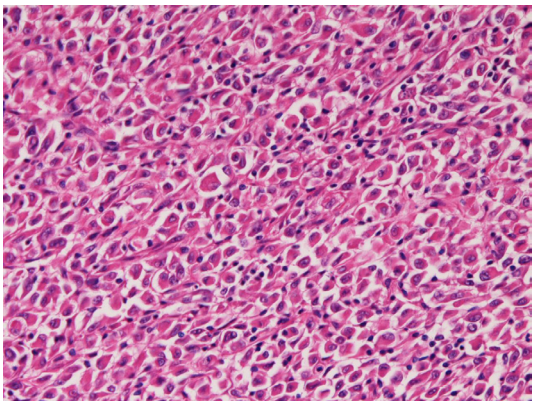

Figure 3. The carcinoma cells show nuclear atypia and rhabdoid features. HE, x200.

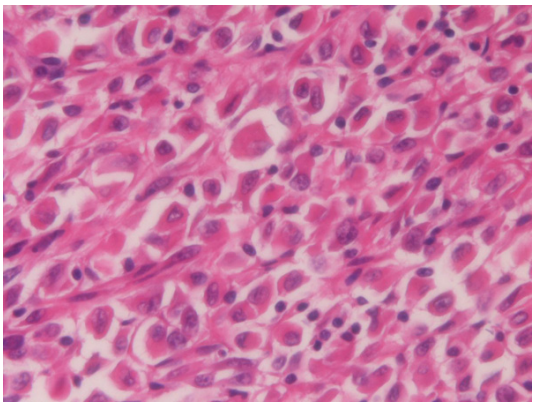

Figure 4. The rhabdoid features and malignant character are apparent. HE, x400.

Discussion

The pathological diagnosis of the present case is very difficult. The cellular atypia and high Ki-67 labeling (40%) indicate that the pelvic tumors are malignant. The acidophilic ample cytoplasm of the tumor cells showed "rhabdoid" features. The present tumors were immunohistochemically negative for mesencymal antigens, myoglobin, myogenin, lymphoma antigens, and neuroendocrine antigens. vimentin. These observations indicate that the present pelvic tumors are not sarcomas.

The present pelvic tumor consisted only of malignant rhabdoid cells. Rhabdoid features

Table 1. Immunohistochemical reagents and results

Antigens	Antibodies (clone)	Sources	Results
Pancytokeratin	AE1/3	Dako Corp. Glostrup, Denmark	-
Pancytokeratin	polyclonal wide	Dako	-
Pancytokeratin	KL-1	Immunotech, Marseille, France	-
Pancytokeratin	CAM5.2	Becton Dickinson Co. CA, USA	-
HMWCK	34βE12	Dako	-
CK5/6	D5/16	Dako	-
CK7	N1626	Dako	-
CK8	35βH11	Dako	-
CK14	LL002	Novocastra, Newcastle upon type, UK	-
CK 18	DC10	Dako	-
CK 19	RCK 108	Progen, Heidelberg, Germany	-
CK20	K20.8	Dako	-
Melanosome	HMB45	Dako	-
EMA	E29	Dako	-
Vimentin	Vim 3B4	Dako	+++
CEA	polyclonal	Dako	-
Desmin	D33	Dako	-
S100 protein	polyclonal	Dako	-
ASMA	1A4	Dako	-
Myoglobin	polyclonal	Dako	-
Myogenin	polyclonal	Santa Cruz	-
CD34	NU-4A1	Nichirei, Tokyo, Japn	-
p53 protein	DO-7	Dako	-
p63	4A4	Dako	-
Ki-67	MIB-I	Dako	40%
CD3	PC3/188A	Dako	-
CD20	L26	Dako	-
CD30	BERH2	Dako	-
CD45	PD7/26	Dako	-
CD45RO	UCHL1	Dako	-
Chromogranin	DAK-A3	Dako	-
Synaptophysin	Polyconal	Dako	-
NSE	BBS/NC/VI-H14	Dako	+
CD56	UJ13A	Dako	-
CD68	KP-1	Dako	-
KIT	polyclonal	Dako	-
PDGFRA	polyclonal	Santa Cruz, CA, USA	+

+++, 67-100% positive. ++, 33-67%. +, 1-33% positive. -, negative. HMWCK, high molecular weight cytokeratin. CK, cytokeratin. EMA, epithelial membrane antigen. CEA, carcinoembryonic antigen. ASMA, α-smooth muscle antigen. NSE, neuron-specific enolase. PDGFRA, platelet-derived growth factor receptot-α.

ocan occur in any tumors. A few reports of tumors with rhabdoid features have been reported in the renal pelvis [4-6]. However, these reported cases showed areas of urothelial carcinoma in the tumor, and expressed CKs. The term of rhabdoid tumor is confusing. Generally, it is used in a pediatric kidney malignancy called rhabdoid tumor [7, 12]. The kidney rhabdoid tumor is characterized by cytoplasmic filamentous inclusions, prominent nucleoli, duel differentiations into both epithelial and mesenchymal phenotypes, and highly aggressive biological behaviors. Vimentin and CK are positive in these tumors. In the present study, there were no cytoplasmic filamentous inclusions and nucleoli are not prominent. In addition, the present patient is middle-aged man and the pelvic tumors were not so aggressive. Furthermore, the present pelvic tumors showed positive vimentin and negative CKs. These observations indicate that the present pelvic tumors are not the pediatric rhabdoid tumor of the kidney.

Several cases of sarcomatoid urothelial carcinoma have been reported [2, 3]. Sarcomatoid urothelial carcinomas with rhabdoid features have also been reported [4, 5]. In the present pelvic tumors, there were no urothelial carcinoma elements. In addition, the present pelvic tumors were immunohistochemically positive for only vimentin, and were negative for epithelial markers. As is well known, vimentin is not a specific marker for mesenchymal cells, and it is expressed in epithelial tumors.

The author speculates that the present pelvic malignant 'rhabdoid' tumors are not sarcomas

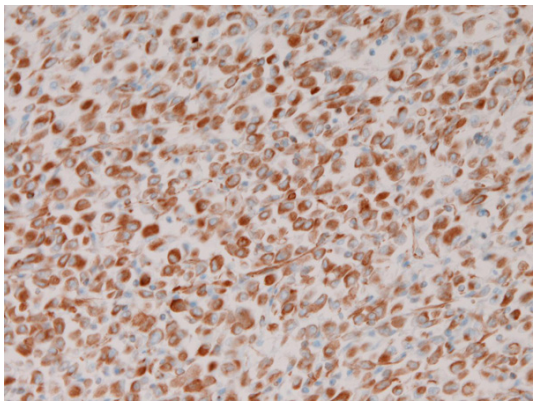

Figure 5. The tumor cells are strongly positive for vimentin. Immunostaining, x200.

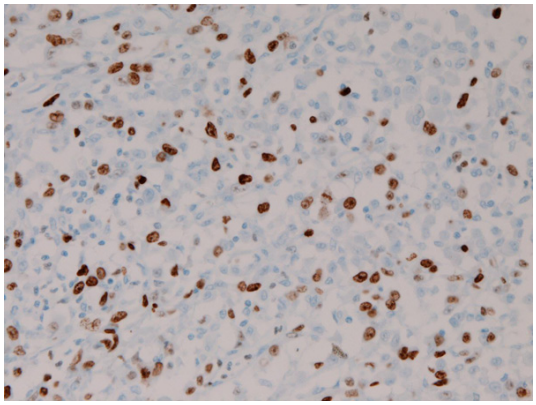

Figure 6. The tumor cells shows Ki-67 antigen (labeling 40%), Immunostaining, x200.

or rhabdoid tumors but urothelial carcinomas composed only of rhabdoid cells with complete loss of CKs. The mechanism of the loss of CKs is unknown. The multiple tumors in the renal pelvis and multiple papillary urothelial carcinomas of the urinary bladder in the present case suggest the above statement.

Conflict of interest statement

The author declares that he does not have any competing interests.

Address correspondence to: Dr. Tadashi Terada, Department of Pathology, Shizuoka City Shimizu Hospital, Miyakami 1231 Shimizu-Ku, Shizuoka 424-8636, Japan. Tel: 81-54-336-1111; Fax: 81-54-336-1315; E-mail: piyo0111jp@yahoo.co.jp

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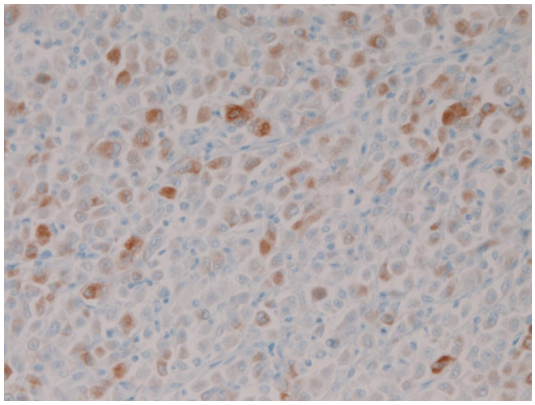

Figure 7. The tumor cells are focally for neuron-specific enolase. Immunostaining, x200.

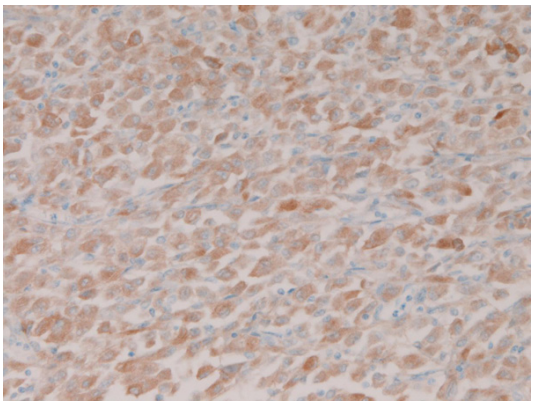

Figure 8. The tumor cells are focally positive for platelet-derived growth factor receptor-α, but this is cytoplasmic staining. Immunostaining, x200.

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