A CASE OF MALIGNANT RENAL NEOPLASM WITH RHABDOID FEATURES IN AN ADULT

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Abstract : We report a case of malignant neoplasm with rhabdoid features of the kidney in a 68 year old man. To our knowledge malignant rhabdoid tumor of the kidney (MRTK) has been limited to the pediatric age group. Malignant renal neoplasm similar to MRTK occurring in adults is very rare. However, there has been a recent review in which the authors found 23/480 cases of renal cell carcinoma (RCC) (4.7%) exhibiting rhabdoid features, all in adults. In the present case, examination of many sections of tumor-affected tissue revealed no sign of any other malignant tumor included RCC, so-called pure adult malignant rhabdoid tumor in the kidney. MRTK does not respond well to aggressive chemotherapy regimens and survival tends to be short. This patient remains alive with no metastasis and local recurrence more than 45 months from the operation without adjuvant therapy .

Key words: rhabdoid tumor, malignant renal neoplasm, adult

INTRODUCTION

Malignant rhabdoid tumor of the kidney (MRTK) is a rare renal tumor observed in infants. Patients with this tumor have a very poor prognosis. We recently encountered an adult with renal tumor resembling MRTK in terms of histological features, as presented in this paper.

CASE REPORT

Abdominal ultrasonography, performed during thorough health checks, revealed a tumor in the left kidney of a 67-year-old Japanese male. He consulted our department, where physical examination of his thoracic and abdominal areas and blood biochemistry revealed no abnormalities. However, abdominal CT scans indicated a tumor with a major diameter of 6 cm in the left kidney, and contrast enhancement in some parts of the tumor (probably the parts representing tumorous vessels) (Fig.1). No signs of metastasis were evident on thoracic and abdominal CT or bone scintigraphy. The patient was thus suspected of having renal cell carcinoma and transabdominal radical nephrectomy was performed on October 3, 1997. Macroscopically, the tumor had affected the entire inferior pole of the kidney and appeared to be solid and composed of many yellowish white nodes. Hemorrhagic necrosis was seen in some areas. Histopathological examination of the specimen stained with H&E revealed spindle-oval tumor cells arranged in a haphazard pattern with many rhabdoid cell-like tumor cells whose cytoplasm contained an eosinophilic globular configuration and

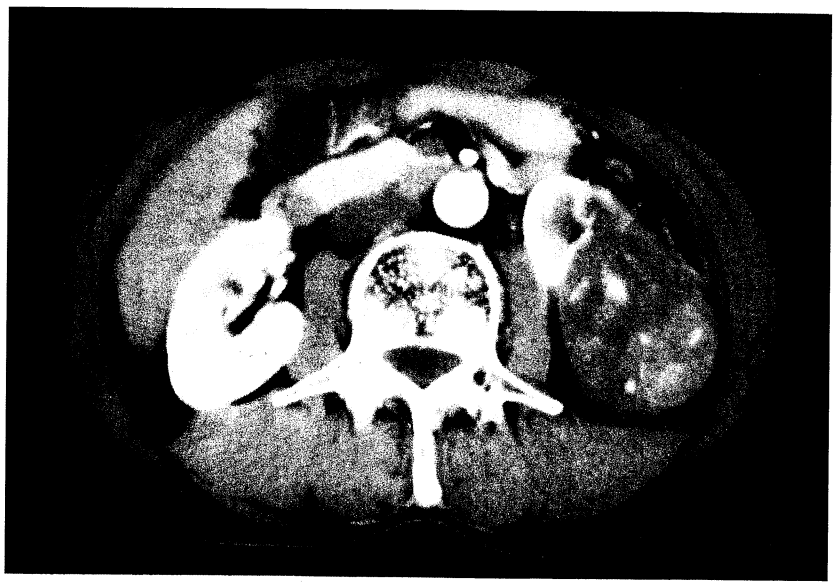

Fig. 1. CT of abdomen
CT scan shows 6 cm mass with contrast enhancement in some parts of the tumor.

Fig. 2. Microscopic study (HE stain × 400)
Microscopic study shows rhabdoid cell-like tumor cells whose cytoplasm contained an eosinophilic globular configuration and prominent nucleoli.

prominent nucleoli (Fig.2). The immunohistochemical findings showed all negative for keratin, EMA, desmin, SMA and S-100 protein, and positive only for vimentin (Fig. 3). No other malignant tumor was found in any other sites. The patient was diagnosed as having non-epithelial malignant tumor with rhabdoid features. No adjuvant therapy was

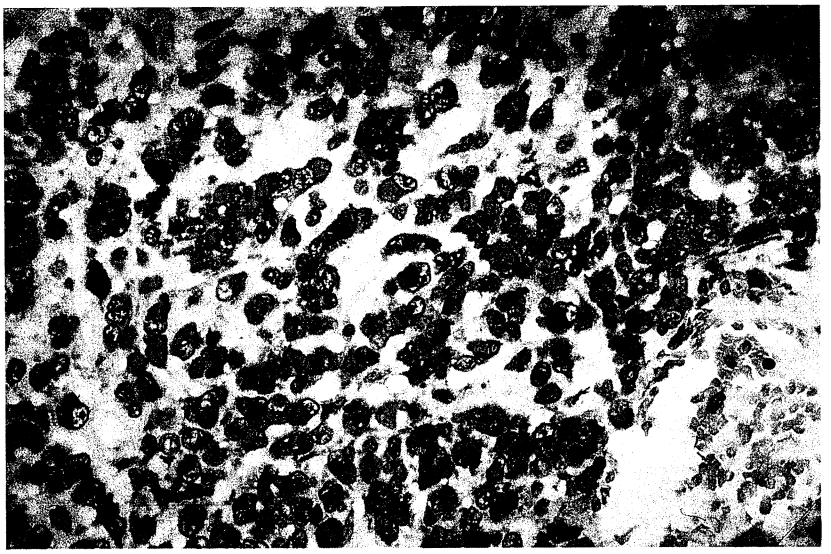

Fig. 3. Microscopic study (immunohistochemical stain × 400)
Microscopic study shows positivity for vimentin.

administered after surgery. At present, 45 months after surgery, the patient shows no evidence of disease (NED).

DISCUSSION

In 1978, Beckwith et al.[1] classified MRTK as a sarcomatous subtype of Wilms' tumor. Later, MRTK came to be viewed as a malignant tumor different from Wilms' tumor, based on reports of cases with this tumor in the various organs and its immunohistochemical and electron microscopic features. Even in infants, MRTK is a very rare disease. According to the NWTS (National Wilms' Tumor Study)[2] prevalence covering the period until 1989, MRTK accounted for only 1.8% of all renal tumors in children. Also, when clinical features are compared the peak age upon detection differs between MRTK (13 months on average) and the favorable histology (FH) type of Wilms' tumor (2-3 years)[3]. Furthermore, according to an NWTSII report[4], the disease-free two-year survival rate differs between the FH type Wilms' tumor (90%) and unfavorable histology (UF) type Wilms' tumor including MRTK (54%). Of the cases of MRTK adequately followed up, 80% died, indicating that patients with MRTK have a quite poor prognosis[2]. At present, this tumor is viewed as an independent disease entity within the category of pediatric renal tumor[2, 5, 6].

Previous reports have concerned 6 adults with renal tumor, accompanied by malignant rhabdoid tumor (MRT)-like histological features. Of these 6 cases, 1 was reported as a case of renal cell carcinoma (RCC) complicated by MRT[6] and 2 were reported as transitional cell carcinoma complicated by MRT[7, 9]. The number of cases of MRT-like renal tumor reported, without complication by any other malignant tumor like the case presented in this paper, so-called pure adult malignant rhabdoid tumor in the kidney, is four including the present case

Table 1. Reported cases of pure adult malignant rhabdoid tumor in the kidney

References	Age (years)	Sex	Tumor size (cm)	Metastasis	Treatment	Prognosis
Lowe W[10]	32	female	12	none	nephrectomy	PD
Ebbinghaus SW[11]	56	male	9.0	lung	chemotherapy	PR
Clausen HV[12]	54	female	18	lung	chemotherapy	PD (died 3 months following admission)
The present case	68	male	6.0	none	nephrectomy	CR (45 months no evidence of disease)

[10, 11, 12] (summarized in table 1). Recently, however, Neriman et al. reported that rhabdoid cells were observed in 4.7% of cases in a large consecutive series of 480 patients with RCC, and the rhabdoid component was always admixed with usual RCC[13]. In the present case, examination of many sections of the tumor-affected tissue revealed no sign of any other malignant tumor. Histological features characteristic of MRT are: 1) the presence of rhabdoid cells which resemble striated muscles cells and possess esinophilic globular configuration in the cell body; 2) negative immunostains for myogenic enzymes such as myoglobin; 3) positive immunostains for vimentin and cytokeratin[14]; 4) absence of differentiation into ultrafine structure or myoblasts (a known change of rhabdomyosarcoma) under an electron microscope; and 5) the finding of filamentous structures in the globular configuration[5]. All these features, except for negative immunostain for cytokeratin, were observed in the present case.

Because no disease entity suitable to express the histological features of this adult case was available, we diagnosed this patient as having non-epithelial malignant tumor with rhabdoid features. According to the criteria proposed by Dr. F.K. Mostofi of the Armed Forces Institute of Pathology (AFIP), this case can be pathologically rated as having unclassified malignant neoplasm with rhabdoid features. Generally, MRTK is a highly therapy-resistant malignant tumor with a very poor prognosis[15]. Two adults, reported as having MRTK-like features, showed rapid progression of their disease and died[10, 12]. In the other adult with MRTK-like features, the tumor metastasized to the lungs and resisted various therapies, but the lung metastasis disappeared following interleukin II therapy. This patient has survived for 18 months[11]. The patient presented in this paper received no adjuvant therapy after radial nephrectomy and is free of local recurrence and metastasis at present, 45 months after surgery. This case needs to be followed up carefully.

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