

## Sarcomatoid Renal Cell Carcinoma with an Unusually Rapid Fatal Course

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**= Abstract = Sarcomatoid renal cell carcinoma is an uncommon neoplasm consisting of typical renal cell carcinoma associated intimately with a sarcomatoid component. The prognosis is known to be very poor. We present an autopsy case of sarcomatoid renal cell carcinoma that went through a highly fulminant lethal course. The patient was a 62-year-old male with left flank pain and weight loss. Computed tomograph and aortogram revealed a left renal mass growing out into the retroperitoneum. Multiple metastatic nodules were detected in the lungs and bone. Histologically the tumor showed features of anaplastic carcinoma, malignant fibrous histiocytoma and fibrosarcoma. Immunohistochemical studies of tumor revealed positive staining for cytokeratin (CAM 5.2). We interpreted this tumor as a sarcomatoid renal cell carcinoma extending into the retroperitoneum. He died of respiratory failure secondary to massive lung and mediastinal involvement of the tumor masses.**

Key Words: *Renal cell carcinoma, Sarcomatoid carcinoma, Kidney*

### INTRODUCTION

Sarcomatoid renal cell carcinoma has been recognized as a distinct renal tumor since it was first named by Farrow *et al.* in 1968. It is an uncommon, but not rare, neoplasm consisting of typical renal cell carcinoma associated intimately with a sarcomatoid component. Recognizing this entity is important because it is highly lethal, and histologically, the sarcomatous component may mimic almost any sarcoma, including fibrosarcoma, malignant fibrous histiocytoma,

leiomyosarcoma, and osteosarcoma. In most cases, however, major sarcomatoid patterns are malignant fibrous histiocytoma and fibrosarcoma (Ro *et al.* 1987).

Although the prognosis of sarcomatoid renal cell carcinoma is known to be extremely poor (Farrow *et al.* 1968; Tomera *et al.* 1983), little is known about the clinicopathologic parameter of prognostic significance. In M.D. Anderson Cancer Center series, the two and three year survival rates were reported to be 28.6 and 19.0 per cent, respectively (Ro *et al.* 1987). They found the stage of the disease and tumor necrosis in the sarcomatoid area to be significant predictors of the outcome. We hereby present a case of sarcomatoid renal cell carcinoma that

went through a rapid lethal course.

## CASE REPORT

This 62-year-old male had been healthy except for a mild discomfort of his left flank for 2 years before admission to Seoul National University Hospital on July 1992. He lost 5 kg of his body weight in recent several months. Cough and sputum developed from three weeks before admission. The symptoms aggravated at night and were not related to the meal. The flank pain was of a dull nonradiating character and was aggravated by coughing and was relieved by left lateral decubitus position. Past medical history included tuberculous pleurisy at 26 years of age, and acute myocardial infarction 6 years before admission. At admission, physical examination showed systolic murmur at the left sternal border and ill-defined mass in the left upper quadrant of his abdomen. The mass was firm and had a smooth surface. A mild tenderness was noted. Rest of the physical examination was normal. Laboratory findings showed : white blood cell count  $9200/\text{mm}^3$ , hemoglobin 14.2 gm/dl, and platelet  $355,000/\text{mm}^3$ , blood urea nitrogen 19 mg/dl, creatinine 1.1 mg/dl, uric acid 3.9 mg/dl. Liver function tests were normal. Urinalysis showed trace albumin, and 1-4 red blood cells per a high power field. Chest and abdomen radiographs revealed multiple nodules in the lung, pleural effusion and mass shadow in the abdomen. Gastrofiberscopy revealed compression at the fundus of stomach by an extrinsic mass. Computed tomograph of the abdomen disclosed a mass in the left kidney extending to the retroperitoneum. Bone scan revealed spots of hot uptake in the left ilium and sacroiliac joint. Needle biopsy of the kidney mass revealed features suggestive of malignant fibrous histiocytoma. Since the tumor mass revealed vascular supply from the renal artery, therapeutic embolization through the renal artery was once tried. Over the subsequent few weeks he developed multiple metastases, pleural effusion and massive ascites although chemotherapy with

vincristine, melphalan and prednisolone was started. Pleurodesis was performed using picibanil. Antibiotics were given for control a suspicious bacterial infection of *Corynebacterium* species, detected by a blood culture. He went through progressive downhill course and died of respiratory failure 34 days after admission.

Postmortem examination showed a mildly icteric body with peripheral edema and abdominal distention. The thoracic cavity was filled with massive gray white tumor masses involving pleura, hilar lymph nodes, pericardium and diaphragm (Fig. 1). Two liters of serosanguineous effusion was noted in the thorax. Multiple nodules were also seen in both lung parenchyma. The abdominal cavity contained serosanguineous ascites. There was a lobulated firm mass in the left kidney. The tumor was fixed firmly to the retroperitoneum. The tumor mea-



Fig. 1. Left thoracic content showing massive gray white tumor masses in the pericardium(P) and diaphragm(D).

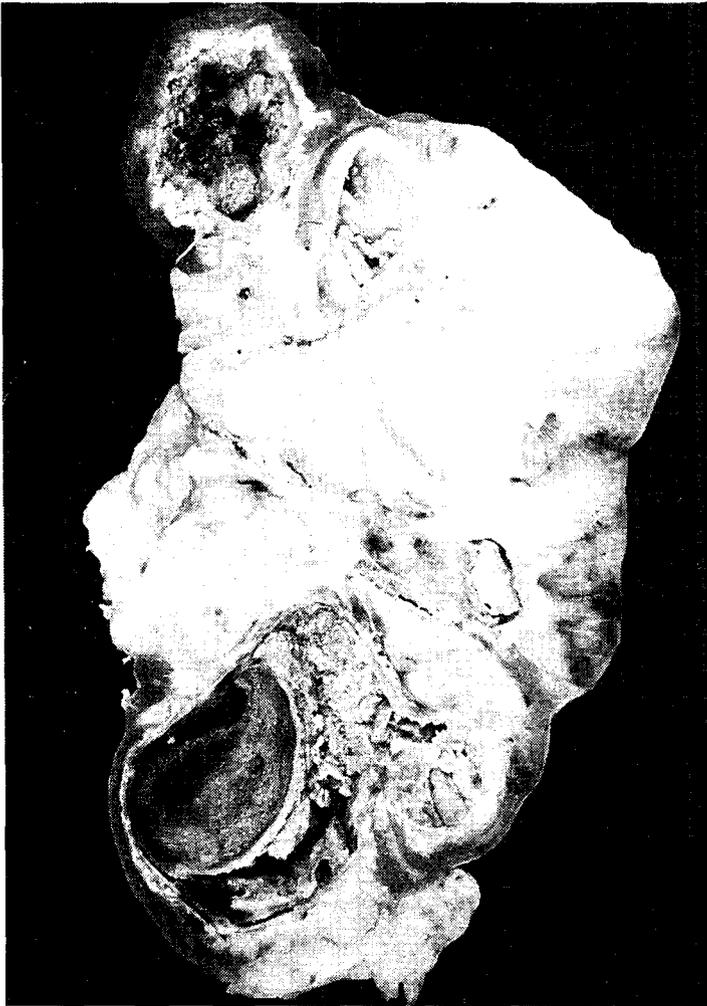


Fig. 2. Sagittal section of the left kidney shows irregular gray white lobulated mass involving the entire thickness of the kidney. Two large areas of necrosis(chemoembolization effect) are seen in both poles.

sured 14 x 10 x 6 cm. Cut sections of the kidney and the mass disclosed gray-white fleshy mass in the kidney, extending toward retroperitoneum. Main portion of the mass was in the retroperitoneum. However, this mass was apparently a bulged portion of the intra-renal mass and broke through the Gerota's fascia(Fig. 2). Yellowish necrosis was seen in one third of the intra-renal tumor, which was thought to be ischemic necrosis secondary to renal artery chemoembolization. The liver, spleen, pancreas were sites of multiple metastasis and vertebral body was also involved(Fig. 3). Multiple lymph nodes in the thoracic and abdominal cavity were involved by the tumor.

Microscopically, there were two cytologically different portions, one with pleomorphic cells

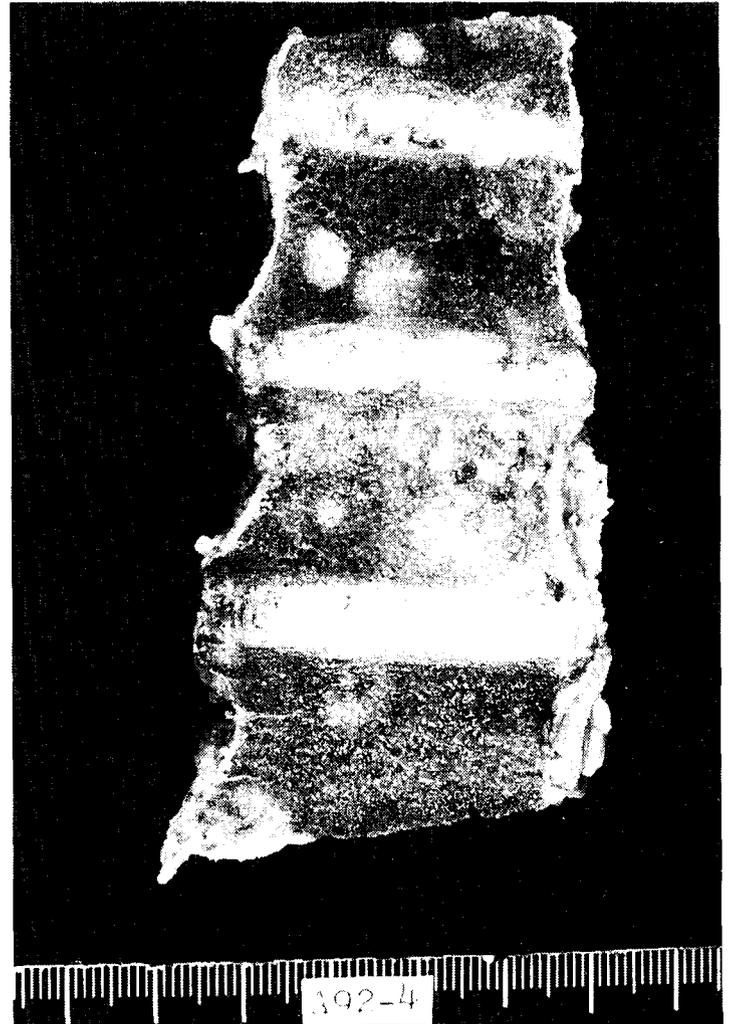


Fig. 3. Metastatic nodules in the vertebra.

and the other with spindle cells. The pleomorphic portion showed numerous multinucleated giant cells and many mitoses(Fig. 4). The cytoplasm of these pleomorphic cells was abundant and deeply eosinophilic. Periodic acid Schiff reaction and oil red O stain failed to reveal any glycogen or lipid droplets in these cells. This portion was reminiscent of anaplastic carcinoma or malignant fibrous histiocytoma. Meanwhile, the remaining portion was basically spindle cell sarcoma. The cellular arrangement was of fibrosarcoma and there was storiform pattern (Fig. 5). This portion resembled the needle biopsy specimen of this patient. Immunohistochemical studies of tumor from two different portions showed same findings. They revealed positive staining for cytokeratin(CAM 5.2), vimentin, desmin, alpha 1-antitrypsin and alpha 1-antichymotrypsin. The cytokeratin expression was focal but definite in spindle cells. The tumor cells were

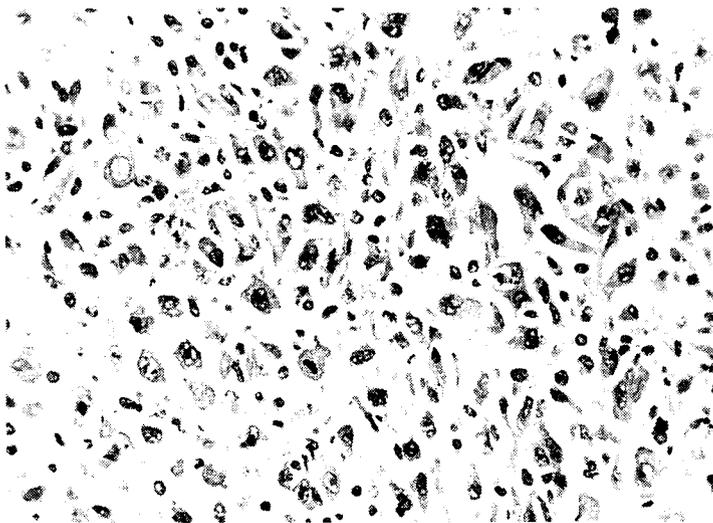


Fig. 4. Photomicrograph of the pleomorphic portion of the tumor.

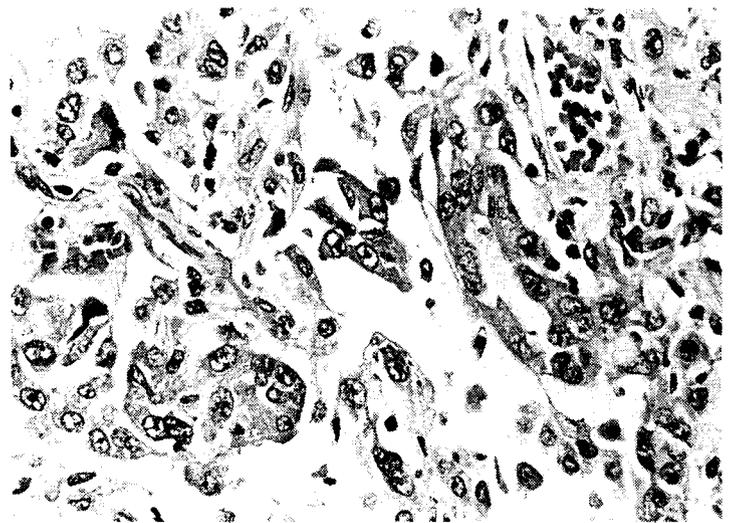


Fig. 6. Photomicrograph of the metastatic lung lesion showing definite glandular elements.

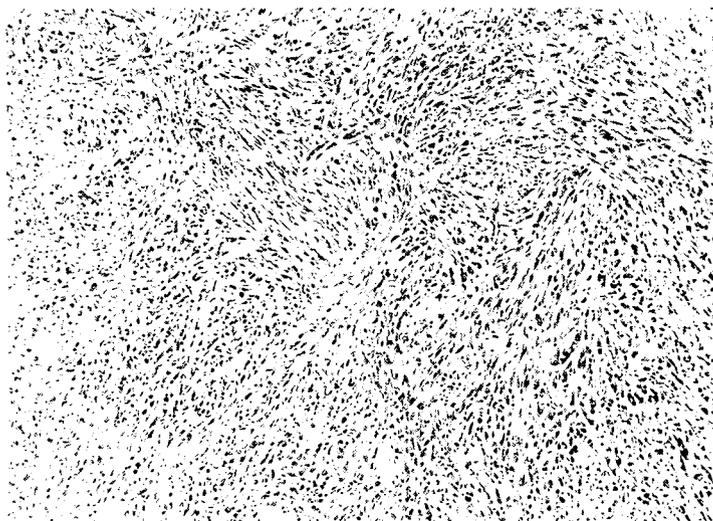


Fig. 5. Microscopic picture of the spindle cell portion showing storiform-like pattern.

negative for myoglobin and S-100 protein. Ultrastructurally, the majority of tumor cells showed features of histiocytes or smooth muscle differentiation. Epithelial character could not be seen. The histology of tumor in the metastatic lesions was similar to the original tumor. However, there was a few areas of glandular differentiation amongst spindle cell growth in the lung (Fig. 6).

Findings of other organs included an old myocardial infarct in the inferior wall of the left ventricle associated with an eccentric atheromatous narrowing at a distal obtuse marginal branch of the left coronary artery. The brain was not examined.

## DISCUSSION

Sarcomatoid renal cell carcinoma is a tumor that is often confused with true sarcoma of the kidney. Tomera *et al.* (1983) have indicated that most tumors that were thought to be primary renal sarcomas were actually sarcomatoid renal cell carcinomas when a careful search was made. In the majority of sarcomatoid renal cell carcinoma, the diagnosis depends on finding evidence for epithelial differentiation (Farrow *et al.* 1968; Ro *et al.* 1987; Bertoni *et al.* 1987; Tomera *et al.* 1983). This epithelial differentiation can be suggested by ultrastructural examination (Bonsib *et al.* 1987; Deitchman and Sidhu 1980) and by immunohistochemistry. Khorsand *et al.* (1986) reported cytokeratin expression of the spindle cell component in 16 out of 28 cases of sarcomatoid renal cell carcinoma. Additionally, cytokeratin immunostaining may highlight a poorly defined epithelial component suspected on routine stains. In our case the tumor had been called malignant fibrous histiocytoma with needle biopsy specimen. Cytokeratin was negative at that time. However, autopsy specimen showed focal cytokeratin positivity as well as glandular structures in the metastatic lung lesion. Electron microscopy was not of help in our case.

Rapid progression of the disease with fatal outcome in this patient is not surprising for this

type of tumor. Tomera *et al.* reported that most patients of sarcomatoid renal cell carcinoma died of metastatic disease within a year after diagnosis with a median postoperative survival of 6.3 months (ranged from 1 to 14 months). When sarcomatoid area is mixed with carcinomatous area in the kidney tumor the percentage of the sarcomatoid component can be a determinant for the prognosis (Ro *et al.* 1987; Bertoni *et al.* 1987). In other words when the sarcomatoid component was less than 5 per cent of the entire tumor masses the prognosis was much better (Bertoni *et al.* 1987). In our case, it was a pure sarcomatoid carcinoma. Another important finding in our case was an extensive lymph node metastases, which are not usual in most cases of sarcomatoid renal cell carcinoma (Bertoni *et al.* 1987).

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