Journal of Korean Medical Science
Vol. 11, No. 4, 351-355 Aug. 1996

Hemangiopericytoma of Renal Sinus Expanding to the Renal Hilum : An Unusual Presentation Causes Misinterpretation as Transitional Cell Carcinoma

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We report a case of renal hemangiopericytoma occuring in renal sinus and expanding to the renal hilum. This unusual presentation caused misinterpretation of this tumor as transitional cell carcinoma of the renal pelvis clinically. The patient who was a 30-year-old woman had a relatively well demarcated solid tumor, 8×6 cm, in the renal sinus of the left kidney.

Key Words : *Hemangiopericytoma, Renal sinus*

INTRODUCTION

The hemangiopericytoma of the kidney is extremely rare. In world literature, only 21 cases of renal hemangiopericytoma have been reported. Although hemangiopericytoma can occur where the capillary and venules are present, renal capsule and parenchyma have been known as a frequent location of the tumor in the kidney(Heppe et al.,1991). There was no previous report of renal hemangiopericytoma which occured in the renal sinus masquerading as a transitional cell carcinoma clinically. We report an unusually presenting hemangiopericytoma in the renal sinus with the review of pathologic and clinical characteristics of the tumor.

CASE REPORT

A 30-year-old woman was admitted to Kangnam St. Mary's Hospital in November 1994 for general weakness and intermittent cold sweating. Physical examination revealed a palpable mass on the left quadrant of the abdomen. His blood pressure was 120/80 mmHg and pulse rate 80/min. Fasting blood sugar level was 84 mg/dl. Laboratory data including hematology, serum electrolytes, liver function test and urinalysis were normal.

A Roentgenogram of the chest was normal. An ultrasonography revealed a well demarcated solid mass in the left kidney. Computed tomography of abdomen showed a well demarcated highly enhanced mass with intravenous contrast medium, which occupied the central portion of the kidney and expanded to the renal hilum(Fig.1). Under the clinical impression of a transitional cell carcinoma of the renal pelvis, total nephrectomy of the left kidney was performed. On the

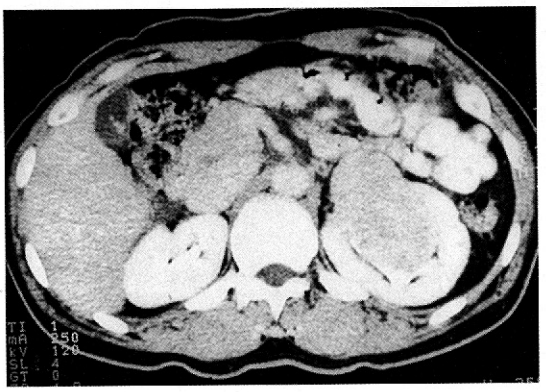

Fig. 1. Computed tomography demonstrates a well demarcated highly enhanced tumor mass in the central portion of left kidney with expanding to the renal hilum.

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operation field, the tumor was markedly adhered to the perihilar areas, but there was no definite invasion to the neighboring structures. The patient was free of tumor for 14 months after the nephrectomy.

PATHOLOGY

Gross examination. The left kidney, which measured 12×9×6cm and weighed 280 gm, had a well demarcated pale yellow to whitish gray ovoid tumor mass(8×6×5cm) in the renal sinus which protruded to the renal hilum(Fig. 2). On cut section, the tumor was solid and firm and contained several nodular areas in the periphery. There were some dilated vascular spaces, small microcysts and focal areas of hemorrhage in the tumor without areas of necrosis. There was markedly collapsed renal pelvis and upward infiltration of the tumor to the upper pole of the renal parenchyma.

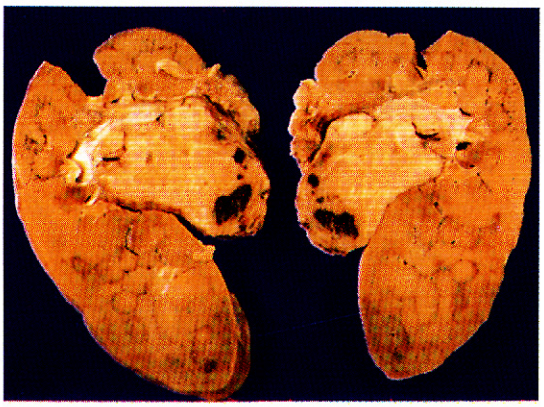

Fig. 2. The tumor, 8×6×5cm, is totally located in the renal sinus with marked collapse of the renal pelvis. There are focal areas of hemorrhage and small microcysts.

Light microscopy. The tumor consisted of mostly solid and hypercellular areas intermingled with some variable sized vascular channels(Fig. 3). There were focal areas of hemorrhage and cystic degeneration accompanied with some aggregation of foamy macrophages. The thin-walled vascular channels, lined by endothelial cells, were mostly compressed by surrounding tumor cells and focally dilated. They had round to oval vesicular nuclei with or without a small inconspicuous nucleolus and a mild to moderate amount of ill-defined cytoplasm. Mitotic activities were rarely found in about one per 30 high power fields.

A few scattered multinucleated giant cells were also found. There was diffuse infiltration by many mononuclear cells and mast cells between the tumor cells. Several nodular areas in the periphery showed marked hyalinization and fibrosis. PAS stained section revealed many collapsed intervening vascular channels which were lined by a single layer of endothelium. Although reliculin fibers were generally sparse, focal areas showed individually wrapped tumor cells by Wilder's reticulin stain(Fig. 4). Except for the extension to the renal hilum and partial invasion into the renal parenchyma by the

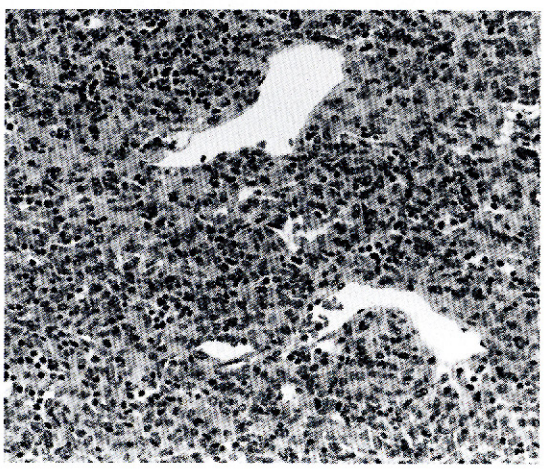

Fig. 3. The tumor shows focally dilated but mostly compressed thin-walled vascular channels by surrounding tumor cells(H & E, ×400).

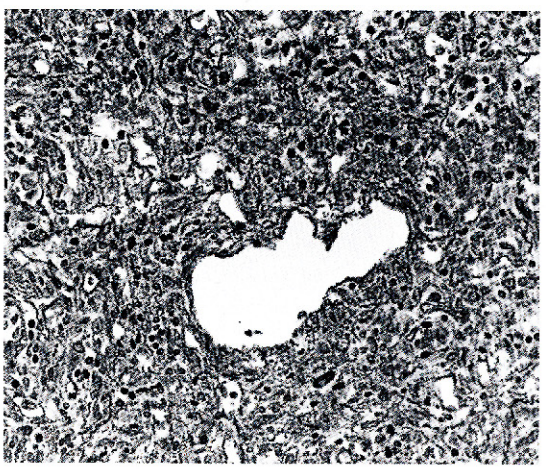

Fig. 4. Reticulin stain reveals generally sparce but focally abundant reticulin fibers between the tumor cells(Wilder's reticulin stain, ×400).

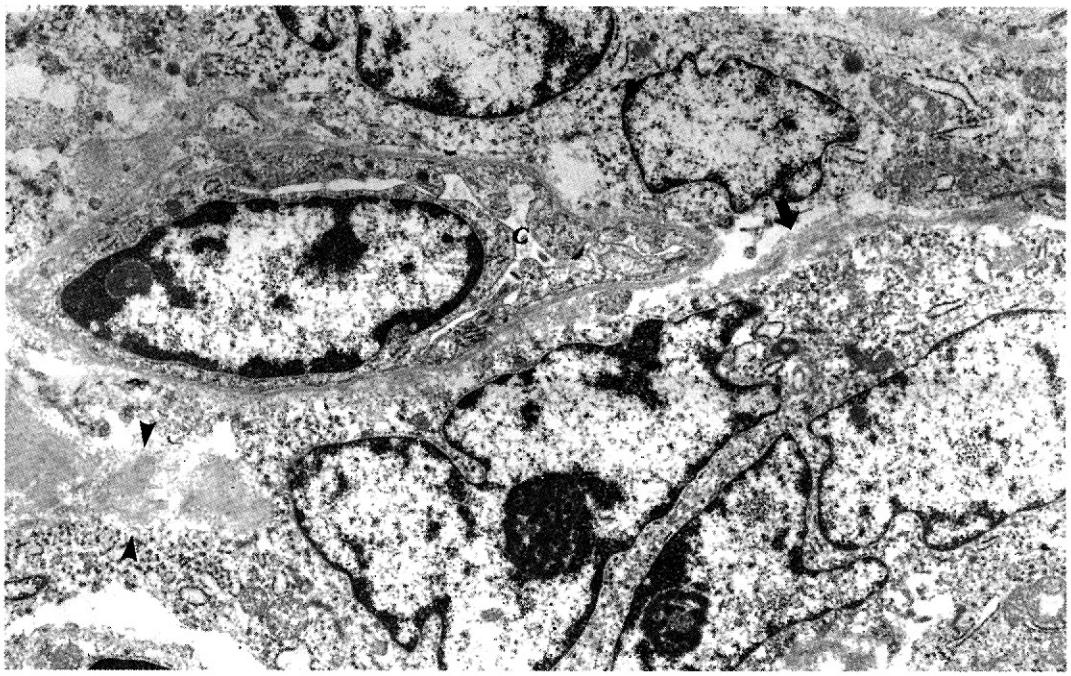

Fig. 5. Electron micrograph demonstrates characteristic relationship between the tumor cell and capillary(C). Discontinuously enveloped basal lamina(arrow) and large amount of basal lamina-like materials(arrow head) are shown. Original magnification× 10,000.

tumor cells, we could not find any invasion into the renal capsule, perirenal tissue, renal pelvis, ureter, renal artery and vein.

Immunohistochemistry revealed positive reaction for vimentin and negative reaction for cytokeratin, alpha-smooth muscle actin, S-100 protein, Factor VIII-related antigen and lysozyme.

Electron microscopy. The tumor cells were ovoid to spindle shaped and relatively well differentiated. They had large ovoid to elongated heterochromatic nuclei with smooth or slightly irregular nuclear membranes and one small to medium sized nucleolus.

The cytoplasm had a small amount of intracytoplasmic organelles, such as rough endoplasmic reticulum with a few free ribosomes, mitochodria, lysosomes, pinocytotic vesicles, and a few bundles of microfilaments. The tumor cells were enveloped by a discontinuous basal lamina and separated from endothelial cells by a distinct single or multi-layered basal lamina. Characteristically there were scattered deposits of large amounts of basal lamina-like materials in the intercellular spaces(Fig. 5).

DISCUSSION

The hemangiopericytoma is a very unusual neoplasm in the kidney. Since Black and Heinemann(1955) reported the first case of renal hemangiopericytoma, only 21 cases have been published in world literature up to the present. Our case is the twenty-second case of renal hemangiopericytoma, and the first reported case occuring in the renal sinus. By a summary of these previously reported cases(Table 1), we found that renal hemangiopericytomas occured in the age ranged from 18 to 68 years(mean 42 years), and female to male ratio was 1.1:1. The presenting symptoms were as follows in descending order : mass, flank pain, hematuria, hypoglycemia(Farrow et al.,1968 ; Asa et al.,1981), hypertension(Weiss et al.,1984 ; Sasaki et al.,1992), and weight loss(Lee et al.,1984). The tumor was 1.5cm to 25cm in diameter, and weighed as much as 3720gm. Among 14 cases in which clinical information was available for the location in the kidney, seven of them were located in the capsule(50%) and the remaining seven were in the renal parenchyma(50%). There was

Table 1. The clinicopathologic summary of previously reported renal hemangiopericytomas

	Reported cases	Present case
Number of case	21	1
Age(yrs)	18-68(mean,42)	30
Sex(M:F)	10:11	Female
Site	capsule(7/14) parenchyme(7/14)	renal sinus
Tumor size	1.5-25cm	8cm
Pseudocapsule	present(12/16) absent(4/16)	absent
Mitosis	absent or few	1/30 HPF
Bilaterality	present(1/21)	absent
Paraneoplastic symptoms	hypoglycemia hypertension	absent
Treatment	surgery	nephrectomy
Follow-up	5 mos-12 yrs	14 mos
Recurrence	1/14	absent
Metastasis	4/21	absent
Mortality	6/14	alive

no previous case report in which a tumor had been developed in the renal sinus.

As our case was located entirely in the renal sinus replacing the renal pelvis to the periphery and expanding to the renal hilum, we thought that it might have originated from pericytes surrounding the vessels of renal sinus. However, we could not exclude the possibility of origination from the renal capsule normally present in this location. The renal hemangiopericytoma is usually unilateral single mass, but could be bilateral. Heppe et al.(1991) reported a case of bilateral hemangiopericytoma with the review of previously reported 19 renal hemangiopericytomas. In their series, a case reported by Robertson et al.(1967) should be excluded because it turned out to be juxtaglomerular cell tumor by the presence of renin granules in the tumor cells. The distinction of juxtaglomerular cell tumor from hemangiopericytoma is important because of the high mortality associated with the latter(Squires et al, 1984). There was frequent encapsulation by pseudocapsule in 12 cases(75%), but not in 4 cases(25%). The mitotic activity was usually absent or infrequent, but as many as 24 mitotic figures per 50 high power fields were found in a case reported by Siniluoto et al.(1988).

The hypoglycemia(Farrow et al.,1968; Asa et al., 1981) and hypertension(Weiss et al., 1984; Sasaki et al, 1992) were known as a commonly associated paraneoplastic symptoms in renal hemangiopericytoma. In general, hypoglycemia was usually resolved spontaneously after the removal of the tumor, but there was a patient who died of hypoglycemic coma due to a few postoperative recurrent episodes(Farrow et al., 1968). The excessive storage of glucose in the tumor cells, which was thought to be a pathogenetic mechanism of hypoglycemia, was confirmed by light and electron microscopy by some researchers(Paullada et al.,1968; Asa et al.,1981).

The treatment of choice is complete excision of the tumor mass. Radiation or chemotherapy has not been generally accepted as an effective adjunct to treatment (Lee and Kay, 1962; McMaster et al., 1975; Heppe et al.,1991).

The prognosis of renal hemangiopericytoma is not so good. Fourteen cases of renal hemangiopericytoma with available follow-up information from 5 months to 12 years(mean 34 months), showed a 7.1% recurrence rate (1/14) and 43% of mortality rate(6/14).

Among six patients who have died, two of them died from distant metastsis, and one from hypoglycemic coma with abdominal recurrence(Farrow et al., 1968; Ordonez et al.,1982). The remaining three patients were simply described without enough explanation that they died of the tumor(Farrow et al.,1968). The interval from initial diagnosis to death was 5 months to 10 years. There have been four cases of renal hemangiopericytoma causing metastasis to distant organs, such as lung, bone and pancreas with a metastatic rate of 18%(4/21). Three of them showed lung metastasis accompanied with invasion to the renal vein and inferior vena cava, and/or bone metastasis(Farrow et al.,1968; Ordonez et al.,1982; Lee et al.,1984). The remaining one had pancreatic metastasis(Heppe et al.,1991). Unfortunately, hemangiopericytoma has been regarded as an unpredictable tumor. Neither tumor size nor encapsulation have proved to be of prognostic significance(Heppe et al., 1991). And the clinical behavior of them cannot always be correlated with the microscopic appearance.

Even so, several microscopic findings, such as increased number of mitotic figures, hypercellularity, cellular pleomorphism, hemorrage and necrosis are implicated as helpful factors in predicting the clinical behavior(McMaster et al., 1974).

During the 14 months follow-up period, our patient has been well and there is no evidence of recurrence or metastasis of the tumor.

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