Metastatic renal hemangiopericytoma: A rare case report

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SUMMARY

The hemangiopericytoma (HPC) of the kidney is extremely rare perivascular neoplasm. There are no specific radiological or clinical findings that can aid in preoperative diagnosis. In the world literature, only 41 cases of renal HPC have been previously documented. We report on a 56-year old woman with renal HPC of the right kidney who had lung metastasis at the time of diagnosis. The right radical nephrectomy was performed and the combination chemotherapy was given, postoperatively. After 3 cycles of chemotherapy, disease was stable, but progression was seen at the end of 6 courses of chemotherapy. The patient died due to progression of disease ten months later after the diagnosis. We suggested that for patients present with renal mass, renal HPC as well as the other renal tumors should be considered in the differential diagnosis.

Key Words: *Hemangiopericytoma; Kidney Neoplasms; Neoplasm Metastasis; Antineoplastic Agents; Radiotherapy*

INTRODUCTION

Hemangiopericytoma (HPC) is an unusual perivascular tumor and first case was described by Stout and Murray in 1942 (1). Moreover, it is classified as a soft-tissue vascular tumor originating from pericytes, which occurs commonly in the extremities, pelvis, head and neck, and meninges (2,3). On the other hand, HPC is rarely encountered in urogenital system (4-6). Renal HPC is also extremely rare tumor and only 41 case reports have been previously documented (7). Herein, we described a case of metastatic HPC localized to the right kidney and reviewed the literature.

CASE REPORT

A 56-year-old woman was referred to Department of Urology, with anorexia, fatigue, back pain, and abdominal bloating which started three months ago, in April 2008. She had history of total abdominal hysterectomy and bilateral salpingo-oophorectomy in 2001 due to uterine leiomyoma. She was not taking any medications. Her family history was non-contributory. Physical examination was normal except for right upper and middle quadrant tenderness. All laboratory tests including complete blood count, liver function tests, urea, creatinine, electrolyte levels were within normal range. Abdominal ultrasonography (USG) revealed a 14 cm solid mass in size localized in the middle-inferior region of the right kidney. Abdominopelvic CT scans showed a 15 cm diameter nodular solid lesion in the right kidney that extended to the hemidiaphragm and possible invasion to the right psoas muscle (Figure 1). A thorax CT scan demonstrated multiple metastatic nodular lesions in the upper lobes of the both lungs, biggest of them measured as 19x17 mm in size (Figure 2). MRI finding of brain was normal. The right radical nephrectomy was performed due to renal tumor. The specimen was macroscopically measured 13x10x9 cm diameter and exceeded to capsule. Histopathologic examination of specimens revealed spindle tumoral cell infiltration arising from mesenchymal cells and they were separated by numerous capillaries and (Figure 3A). The immunohistochemical (IHC) stains of tumor cells were positive for vimentin and CD 34, and negative for Bcl-2, CD 10, CD 99, actin, desmin, and pancytokeratine. The mitotic activity was included four mitoses per 10 high-power fields (hpf) (Figure 3B). In addition, capsule and renal pelvis invasion were also detected in specimen. In the light of these pathological findings, the diagnosis of the right renal HPC was made.

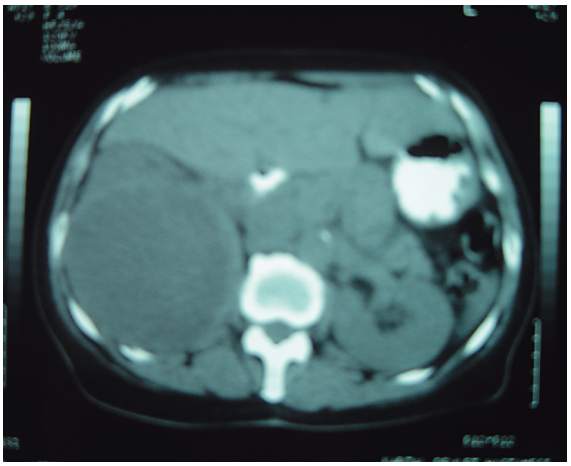

Figure 1. Abdominopelvic CT scans showing a 15 cm diameter nodular solid lesion in the right kidney that extended to the hemidiaphragm and possible invasion to the right psoas muscle

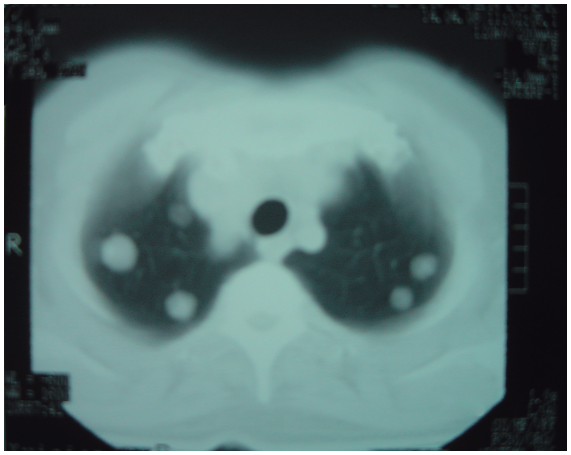

Figure 2. A thorax CT scans demonstrating multiple metastatic nodular lesions in the upper lobes of the both lungs

The patient was admitted to our Department of Medical Oncology with the diagnosis of metastatic renal HPC. The imaging studies showed no residue tumor in the right kidney location except for lung metastasis, post-operatively. After literature

reviewed, the patient was treated with the combination of ifosfamide 1800 mg/m^2, on day 1-5, etoposide 100 mg/m^2 on day 1-5, every three weeks, commutatively with the combination of vincristine 1.4 mg/m^2 on day 1, doxorubicine 75 mg/m^2 on day 1, and cyclophosphamide 1200 mg/m^2 on day 1, every three week. After three cycles of chemotherapy, disease was stable, so six cycles was completed. She tolerated the chemotherapy well, but progressive disease was detected with new liver metastasis and progression in lung metastasis after six cycles. Because of the lack of standard treatment in renal HPC, she was treated with best supportive care during a follow-up of three months. She died due to progression of disease ten months later after the diagnosis.

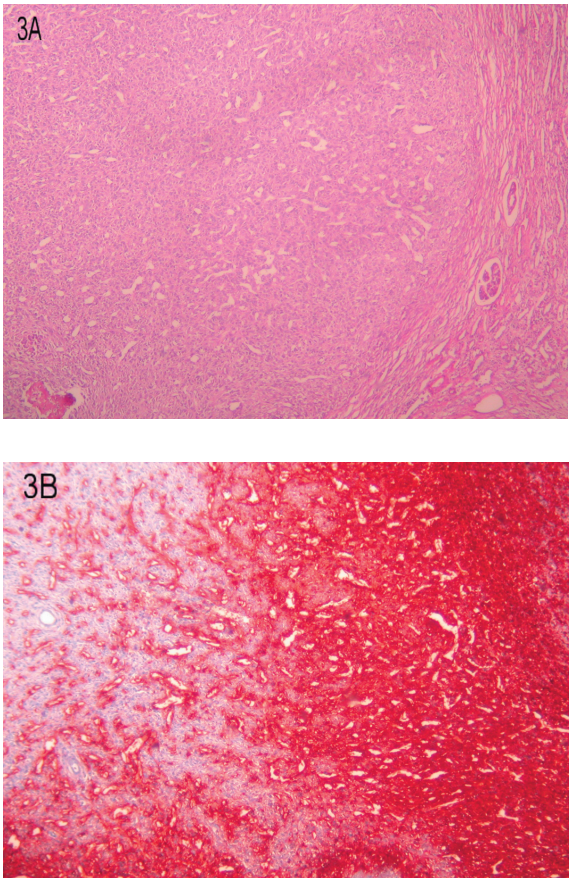

Figure 3. A) Spindle tumor cells separated by numerous capillaries. (H&E, x40), B) CD 34 strongly staining in tumor cells and endothelial cells (x40).

DISCUSSION

HPC is an uncommon soft-tissue vascular tumor that occurs due to the uncontrolled proliferation of pericytes, which are cells spiraling around capillaries (1,2). These cells were firstly described by Zimmerman in 1923 (2). However, the first case of HPC was reported by Stout and Murray in 1942 (1). This neoplasm is very rare in the urogenital system and only 41 cases have been published in the world literature up to this time and two cases of bilateral renal HPC have been reported, previously (7-9).

The mean age of the renal HPC patients at the time of initial diagnosis is 40.3 years (range: 16-68 years). Furthermore, renal HPC patients are slightly younger than other patients with other types of renal cell carcinoma (RCC) (7). There is no difference in the incidence according to gender (10,11). Our patient's age was 56 and compatible with literature. There are no specific signs or symptoms for renal HPC and in 66% of patients a painless mass is the most initial complaint. When 41 cases were analyzed, 19 of them had mass as initial symptom, and the others had pain. Pain in most cases was related to relatively larger masses (7,11). The features of renal HPC cases are summarized in Table 1.

Table 1. Renal hemangiopericytoma patients in the literature

Patient No.	Age/Sex	Initial symptoms	Size (cm)	Treatment	Follow-up	Reference
1	29/F	Hematuria, colic	14x9x9	Surgery	Alive at 1 years	21
2	41/F	Mass	20x15x6	Surgery	ND	22
3	47/F	Mass	10	Surgery	Alive at 11 years	18
4	18/M	Hypoglycemia,mass	13x14	Surgery	ND	23
5	16/M	Hypertension	3	Surgery	ND	13
6	29/F	Pain,mass	8	Surgery+RT+CT	Died at 5 months	24
7	55/M	Pain	7	Surgery+RT	Died at 10 months	24
8	68/M	Pain,mass	16x14x12	Surgery+RT	Died at 2.5 years	24
9	48/M	Pain,mass	10	Surgery+RT	Died at 10 years	24
10	19/M	Hypoglycemia,mass	16	Surgery+RT	Died at 1.5 years	24
11	49/F	Mass	20x10x7	Surgery	ND	25
12	62/F	Hypoglycemia,mass	ND	Surgery	Alive at 1 years	26
13	58/F	ND	2	Surgery	ND	27
14	ND	ND	ND	ND	ND	28
15	40/F	ND	5	Surgery	ND	29
16	20/F	Hypoglycemia,pain	25x21x20	Emb+Surgery	Alive at 3 years	15
17	29/F	Hematuria, cough,pulmonary mass	10x5x5	Surgery+RT+CT	Died at 9 months with metastases	19
18	50/M	Hematuria,mass,pain	18x10x6	Surgery	ND	11
19	56/M	Hematuria, pain	1.5	Surgery	ND	30
20	ND	ND	ND	ND	ND	31
21	ND	ND	ND	ND	ND	32
22	ND/M	Mass	ND	Emb+Surgery	Alive at 1 years	16
23	37/M	ND	5	Surgery	ND	33
24	ND	ND	ND	ND	ND	34
25	33/F	ND	ND	ND	ND	35
26	25/M	Hypertension	4	ND	ND	36
27	39/M	Pain,hematuria (Bilateral)	R 12x10x10, L 13x11x9	Surgery/Surgery	Alive at 3 years with metastases	8
28	27/F	Mass,hypertension	10x9	Surgery	Alive at 18 months	37
29	63/F	Mass	ND	ND	ND	38
30	30/F	Mass	6x6x5	Surgery	Alive at 14 months	39
31	65/F	Mass (incidental)	5x4	Surgery	Alive	40
32	30/M	Mass,hematuria	20x15	Surgery	Alive at 1 years	41
33	51/M	Hypertension	ND	Surgery	ND, remitted hypertension	42
34	21/M	Mass	25x15x10	Surgery	ND	43
35	30/F	Mass	18x10x9	Surgery	ND	43
36	34/F	Mass	3	Surgery	ND	44
37	ND	ND	ND	ND	ND	45
38	61/M	Pain,hematuria	8x10x4	Surgery	Alive at 2 years	46
39	59/M	Abdominal-pulmonary mass	24x17x12	Surgery	Died at 3 months	7
40	37/M	Mass	R 0.4, L 1.5x1.3	Surgery	ND	9
41	43/F	Mass (incidental)	5	Surgery	Alive at 3 months	47
42	56/F	Pain,mass	13x10x9	Surgery+CT	Died at 10 months	Present

ND: no data available, Emb: embolization, RT: radiotherapy, CT: chemotherapy.

Hematuria has been detected in six patients due to the tumor invasion into the pelvicaliceal system, whereas hypoglycemia and hypertension have been noted in 4 and 5 cases, respectively. The cause of hypoglycemia have been associated with the excessive metabolism of the glucose within the tumor (12), however, hypertension is related to a result of renin production from tumors and generally regresses after surgery (13). Our patient was initially presented with back pain and abdominal bloating compared with previous reported cases. We did not detect systemic symptoms, abdominal pain, and hematuria in our patient. However, pulmonary mass as a presenting sign has been noted in 2 cases in the literature. Our patient had also multiple pulmonary metastatic nodules, but she had no pulmonary symptoms related to metastasis.

No specific findings of renal HPC have been found on USG, CT scan or MRI that might aid in the differential diagnosis with the other renal tumors (7). On the other hand, Yaghmai showed that these tumors might have a characteristic pattern in the early arterial phase of angiography, with displacement of the main arteries, presence of the large vessels encircling the tumor, and a well-demarcated tumor stain (14). We also detected no specific signs except for renal mass on USG and CT scan.

The most patients with renal HPC had well-circumscribed or thinly encapsulated tumors, but some are attached to the surrounding tissues. Usually, the size of the tumor is in the range of 1 to 25 cm (Table 1), and in our case, size of the tumor was approximately 14 cm. The amount of intracellular connective tissue is very variable and the diagnosis may be achieved with only the combination of histologic and IHC patterns. These tests are used to exclude other tumors resembling HPC such as malignant fibrous histiocytoma, synovial sarcoma, renal angiosarcoma, and the sarcomatoid RCC (1,3,7). Antibodies against CD 31, CD 34, CD 99, S100, vimentin, cytokeratins, and epithelial membrane antigen are usually used in IHC staining (7). IHC staining of tumor cells revealed positive for vimentin and CD 34, and negative CD 99, pancytokeratine in our patient.

The complete surgical excision was considered to be the cornerstone in first treatment of renal HPC and it was carried out in every reported case. Radical resection is recommended, because incomplete tumor resection associated with consequent higher frequency of relapse and progression (7). Embolization before surgery has been previously reported in only 2 of renal HPC patients (15,16), and they were alive after surgery at 1 and 3 years follow-up.

Chemotherapy (CT) and radiotherapy (RT) have been administered only in adjuvant setting after surgery. Seven patients have been treated with RT, but only one case was alive at 11 years, while the other six survived for a mean of 32 months (17,18). 2 out of these 7 patients treated with RT also received CT, but there was no benefit for survival. They died at 5 and 9 months, respectively (17,19). On the other hand, the benefit of CT has not been demonstrated in the other major series of HPC (3,20). The different schemes including actinomycin D, cyclophosphamide, vicristine, doxorubicine, chlorambucil, methotrexate were used, but produced no favorable results in terms of sustained objective response of metastatic diseases (20). Our patient had metastatic renal HPC. After surgery, she was treated with the CT combination of ifosfamide, etoposide, cyclophosphamide, doxorubicin, vicristine because of soft-tissue vascular tumor. Stable disease was achieved with six cycles, but disease progression occurred after completion of CT. She died 10 months after disease diagnosis.

The prognosis of renal HPC is so poor and the histologic pattern is main prognostic factor. Enzinger and Smith suggested that more mitoses (more than 4 per 10 hpf) were associated with a 29% change of 10-years survival, versus 77% for fewer than 3 mitoses. The other prognostic factors were tumor size and occurrence of necrosis (3). At the time the cases were reported, 12 of 19 patients for whom the follow-up was available were alive, while eight (together with our case) died. In addition, there was evidence of metastasis in 10 patients, but only one was alive (8). The most common site of the metastases is the lungs (7).

In conclusion, renal HPC is an unusual tumor and the only accepted treatment is a wide surgical excision and the efficacy of neither chemotherapy nor radiotherapy has been indicated previously. Its prognosis associates with the size of tumor, age at diagnosis, histological patterns, and recurrence of the tumor. This report also constitutes the 42nd case of renal HPC who had the lung metastases in the literature.

Conflict of interest

We declare no conflicts of interest.

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