Case Report

Renal hemangiopericytoma with lung metastasis: A rare case report

Amitabh Jena, Naru Ramana Reddy, Rashmi Patnayak[1], Amarchala Yadagiri Lakshmi[2]

Departments of Surgical Oncology, [1]Pathology and [2]Radiology, Sri Venkateswara Institute of Medical Sciences, Tirupati, Andhra Pradesh, India

ABSTRACT

Hemangiopericytoma (HPC) is an unusual vascular neoplasm. Renal HPC is very rare. Only a few cases of renal HPC with distant metastasis have been described in the literature. In the absence of specific clinical signs and symptoms and imageological findings, histopathology remains the cornerstone of diagnosis. The present case is a 56-year-old female who presented with abdominal lump and pain of 1-month duration. Her contrast-enhanced computed tomography abdomen and chest showed a large well-defined lobulated heterogeneously enhancing soft tissue density lesion arising from the lower pole of the kidney with central calcification. Nodular opacities were noted in both lungs. It was reported as renal cell carcinoma (RCC) with lung metastasis. She underwent left nephrectomy because of the suspicion of RCC. The histopathology along with immunohistochemistry was reported as HPC. This is an additional case of metastatic renal HPC.

Key words: Hemangiopericytoma, histopathology, lung metastasis, renal tumors

INTRODUCTION

Hemangiopericytoma (HPC) is a vascular neoplasm taking origin from pericytes of Zimmerman. It was first described by Stout and Murray in 1942.[1] It is classified as a soft-tissue vascular tumor originating from pericytes. It occurs commonly in the extremities, pelvis, head and neck, and meninges. It is rarely encountered in urogenital system. Renal HPC is an extremely rare tumor. In world literature, less than fifty cases of renal HPC were reported until 2009.[1] Diagnosis of HPC before surgery is difficult. Almost all the cases were diagnosed after histopathological examination of the excised specimen.[2] The age group of renal HPC patients is between 20 and 50 years.[2] Some patients of renal HPC present with paraneoplastic syndromes such as hypertension, hypoglycemia, electrolyte disorders, and cachexia.[2] We report a case of a 56-year-old female with left renal HPC and with lung metastasis and a brief review of literature.

Address for correspondence: Dr. Amitabh Jena, Department of Surgical oncology, Sri Venkateswara Institute of Medical Sciences, Tirupati - 517 507, Andhra Pradesh, India. E-mail: dramitabh2004@yahoo.co.in

Access this article online

Quick Response Code:

Website: www.ccij-online.org

DOI: 10.4103/2278-0513.197878

CASE REPORT

A 56-year-old female presented with complaints of lump and pain abdomen since 1 month. The clinical impression was renal cell carcinoma (RCC). She was not taking any medications. Her family history was noncontributory. Physical examination was normal except mass in the left loin. All laboratory tests including complete blood count, liver function tests, urea, creatinine, and electrolyte levels were within normal range. Contrast-enhanced computed tomography (CECT) abdomen and chest showed a large well-defined lobulated heterogeneously enhancing soft tissue density lesion measuring 14 cm × 8 cm arising from the lower pole of the kidney with central calcification [Figure 1]. Few nodular opacities were noted in both lungs, largest was in the right lower lobe measuring 1.8 cm × 1.8 cm. The CECT impression was suggestive of RCC left kidney with lung metastasis. The

Cite this article as: Jena A, Reddy NR, Patnayak R, Lakshmi AY. Renal hemangiopericytoma with lung metastasis: A rare case report. Clin Cancer Investig J 2016;5:447-50.

left radical nephrectomy was performed. The cut section of the gross specimen showed compressed adjacent normal-looking renal parenchyma and an ill circumscribed lesion. The lesion was firm and gray-white with nodular appearance [Figure 2]. The histopathology showed compressed renal parenchyma along with a lesion. The lesion comprised spindle cells with elongated nuclei arranged in the form of fascicles. There was moderate pleomorphism and high mitotic activity (8–9/10 HPF). There were focal areas of collagen deposition and presence of foamy cells. Blood vessels were prominent with many blood vessels showing stag horn type of branching pattern [Figure 3]. The tumor was infiltrating the perinephric fat. The ureter was free of tumor. The histopathological possibilities considered were solitary fibrous tumor, HPC, and leiomyosarcoma. A silver stain (Reticulin) highlighted the reticulin fibers around neoplastic cells [Figure 4]. Further immunohistochemistry (IHC) was done with pancytokeratin, vimentin, smooth muscle actin, CD99, and CD34. The positive immunomarkers in the lesional cell were vimentin and CD34. The final diagnosis taking into consideration both histopathological and IHC findings was HPC. The patient recovered well, and he is on adjuvant therapy (combination of methotrexate, doxorubicin, ifosfamide, etoposide, and vincristine).

DISCUSSION

HPC is an unusual vascular neoplasm constituting 2.5% of all soft tissue sarcomas.[3] Black and Heinemann reported the first case of renal HPC in 1955.[4] Less than fifty cases have been reported in world literature so far including a case of bilateral renal HPC.[4] Cases of renal HPC presenting with metastasis are still rarer.[5]

In Indian literature, very few cases of renal HPC have been reported.[6-8] The mean age of the patients is 42 years, suggesting that it occurs at a younger age group compared with RCC.[5] Our patient's age was 56. Renal HPC is slightly more common in females, the ratio of

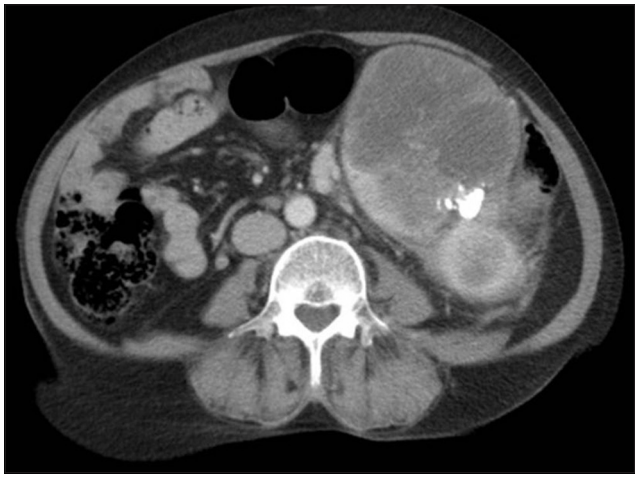

Figure 1: Contrast-enhanced computed tomography showing heterogeneously enhancing mass with calcification from left kidney

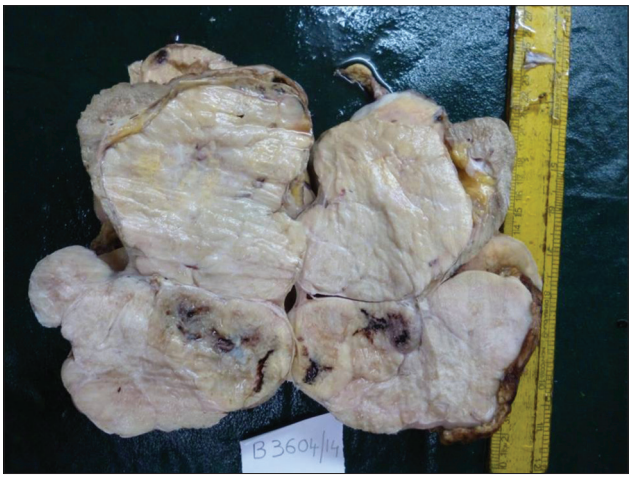

Figure 2: Gross of the gray-white mass with nodular appearance

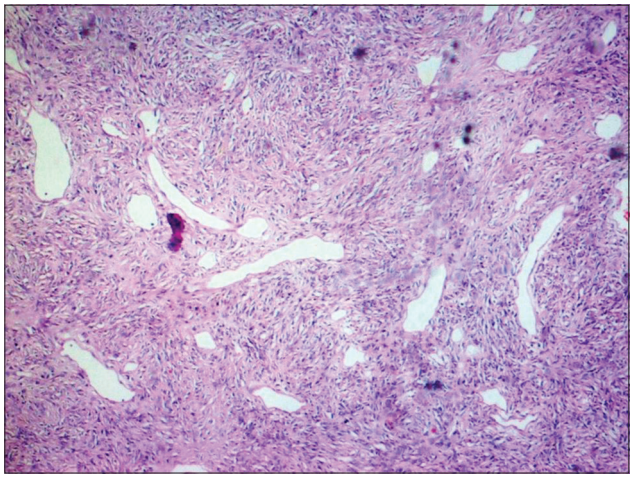

Figure 3: Histopathology showing spindle cell tumor with branched blood vessels (H and E, ×100)

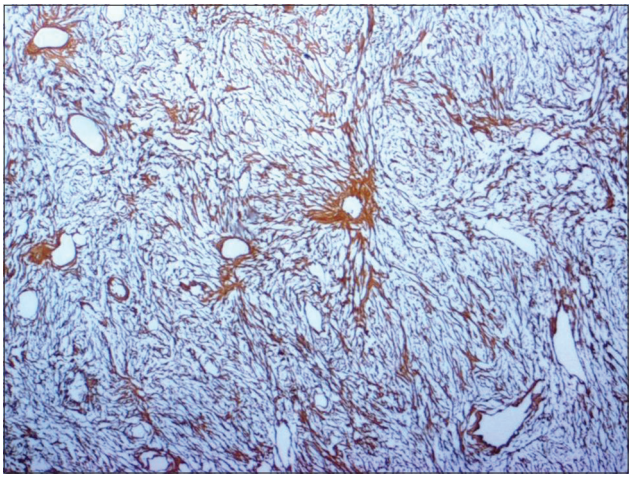

Figure 4: Reticulin stain highlighting the neoplastic cells around reticulin fibers (Reticulin, ×200)

female to male is 1.1:1.[5] There are no specific signs or symptoms. In 66% of cases reported in the literature, initial symptom is abdominal tumor such as noted in our case. Other symptoms in order of frequency are hematuria, hypoglycemia, and arterial hypertension.[5] However, pulmonary mass as a presenting sign has been noted infrequently in the literature.[9] 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 ultrasonography, CT scan, or magnetic resonance image that might aid in the differential diagnosis with the other renal tumors. However, Yagmani reported that these tumors can have a standard characteristic in the early phase of blood angiography of principal arteries, presence of vessels surrounding the tumor and well-marked stain, delimiting the tumor.[10] We also detected no specific signs except for renal mass on CT scan. Macroscopically, most tumors are well-circumscribed lesions or a thin capsule, associated to complex network of vessels and sometimes, with small satellite nodules around the main mass. Only in a few cases, it is adhered to neighboring tissues. The present case showed infiltration to the adjacent tissue with high mitotic activity. The diagnosis of HPC is one of the exclusion. The differential diagnosis includes tumors showing peritheliomatous patterns such as synovial sarcoma and solitary fibrous tumor.[6] Other tumors such as sarcomatoid RCC and leiomyosarcoma also can be considered in the differential diagnosis.

Tumor size reported in the literature range between 2 and 25 cm.[11] In our case, size of the tumor was approximately 18 cm × 10 cm. Main factor associated to prognosis seems to be the histological pattern. Enzinger and Smith observed survival of 77% in 10 years in patients with 0–3 mitoses/10 HPF versus 29% in those patients with four or more by 10 HPF.[12] They also observed the importance of tumor size concerning prognosis. In tumors <6.5 cm, 10 years survival reached 95%, whereas for large tumors, it was 63%.[13] The complete surgical excision was considered to be the cornerstone in the first treatment of renal HPC. Radiotherapy (RT), chemotherapy, or adjuvant or neoadjuvant immunotherapy do not seem to be have a role in the treatment of HPC. In the literature, chemotherapy and 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, whereas the other six survived for a mean of 32 months.[13,14] Two out of these seven patients treated with RT also received chemotherapy, but there was no benefit as far as survival was considered. They died at 5 and 9 months, respectively.[13,15] The benefit of chemotherapy has not been demonstrated in the other major series of HPC.[5,13] In different chemotherapeutic schemes, combinations of actinomycin D, cyclophosphamide, vincristine, doxorubicin, chlorambucil, and methotrexate were used, but there were no favorable results in terms of sustained objective response of metastatic diseases.[15] Our patient had metastatic renal HPC. After surgery, she was treated with the combination of methotrexate, doxorubicin, ifosfamide, etoposide, and vincristine.

CONCLUSION

Renal HPC is an unusual tumor with nonspecific clinical picture and associated imaging findings. The precise diagnosis of HPC is made only after surgery with histopathological examination and relevant IHC. Wide surgical excision remains the treatment of choice. Adjuvant RT, chemotherapy, or immunotherapy has not been found to be effective for renal HPC.

Acknowledgment

The authors wish to thank Professor V. Suresh, for revising the manuscript.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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