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Case Report

Primary hemangioblastoma of the left kidney and epithelioid leiomyoma of the right

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Abstract: Hemangioblastoma is a rare benign tumor; common by found in the central nervous system, about 25% is associated with Von Hippel-Lindau (VHL) disease. Renal hemangioblastoma is extremely rare. The leiomyoma generally occurs in organs with abundant of smooth muscle such as uterus, gastrointestinal tract or subcutaneous soft tissue. It's unusual to see it in the kidney, while the renal epithelioid leiomyoma is even more unusual. Here, we describe a rare case of renal hemangioblastoma and epithelioid leiomyoma occurring in a 43-year-old female without clinical evidence of VHL disease.

Keywords: Hemangioblastoma, epithelioid leiomyoma, renal cell carcinoma, Von Hippel-Lindau disease

Introduction

Hemangioblastoma of the kidney is a rare neoplasm, with uncertain histogenesis. It shares morphological features similar to its cerebellar counterpart [1]. According to report, renal hemangioblastomaoccurs frequently in older people, the average age is 46, no gender difference; it's more likely to be a solitary, isolated solid mass on the right kidney [2-4, 10, 11]. It has similar histological morphological characteristics to renal clear cell carcinoma, and imageological examination often misdiagnosis it as renal cancer [5-7, 12, 13]. Epithelioid leiomyoma is a special type of leiomyoma, and it is extremely rare in the kidney [14, 15]. There are no reliable diagnostic criteria to predict the biological behavior of the tumor [8, 9].

Case report

Clinical history

A 43-year-old female, with no significant systemic disease, who had the left flank pain for more than 20 days was admitted to our hospital for further treatment on April 13, 2016. The patient had neither urinary frequency, urinary urgency, urine pain, hematuria, nor clinical feature indicating the possibility of VHL disease. Physical examination revealed: stable vital signs; right kidney area without knocking pain; no obvious abnormality in laboratory examination. Urinary computed tomography (CT) scan demonstrated a 6.6 × 6.4 × 5.4 cm round heterogeneous density mass in her left kidney with hypodense liquefactive necrosis. The contrast-enhanced CT revealed obvious reinforcement of artery solid components, while low-enhanced in venous phase (**Figure 1F**). A CT scan revealed a round mass with slightly high density in the upper portion of the right kidney, with punctuate calcification inside. The enhancement CT scan showed uneven enhancing with marked no dularenhanced area in the middle (**Figure 2F**). CT diagnosis: 1. The left kidney mass, consider renal cancer; 2. The neoplasm in the upper pole of the right kidney, consider a hamartoma, renal cancer not excluded. On April 19, 2016, the patient underwent laparoscopic right heminephrectomy with general anesthesia, soon after the patient's recovery, performed the left radical nephrectomy on April 26, 2016. After operation, samples were sent for pathological examinations.

Pathological findings

Gross features

The left renal specimen was 12 × 7 × 5 cm in size. In the subcapsular upper pole of the left kidney, there was a defined, yellowish to red, soft to medium texture tumor measuring 5 cm in diameter, with no capsule.

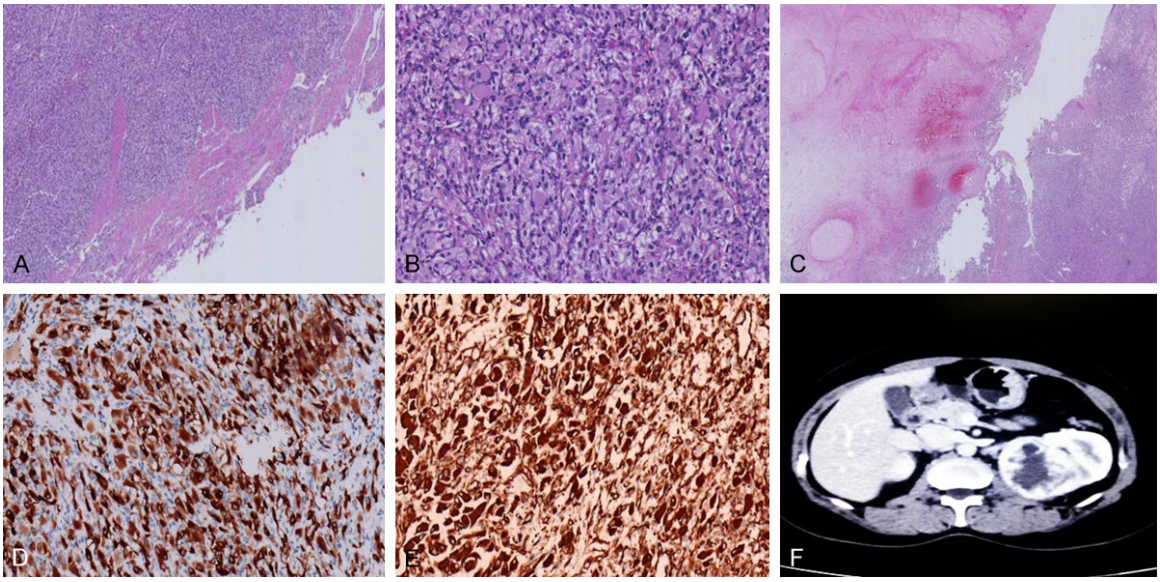

Figure 1. A. The tumor was well-demarcated surrounded by a fibrous psuedocapsule HE × 40; B. The tumor cells showed a rich eosinophilic cytoplasm and nucleus of mediumsize exhibiting occasional lipid droplets and big bizarre nuclei, nuclear fission not having been seen HE × 200; C. Surrounding the tumor we can see a large aera of area-hemorrhage and necrosis HE × 10; D. Immunohstochemical findings: a-inhibin(+) EnVision × 200; E. Immunohistochemical findings: Vimentin(+) EnVision × 200; F. Contrast-enhanced CT scan of left kidney.

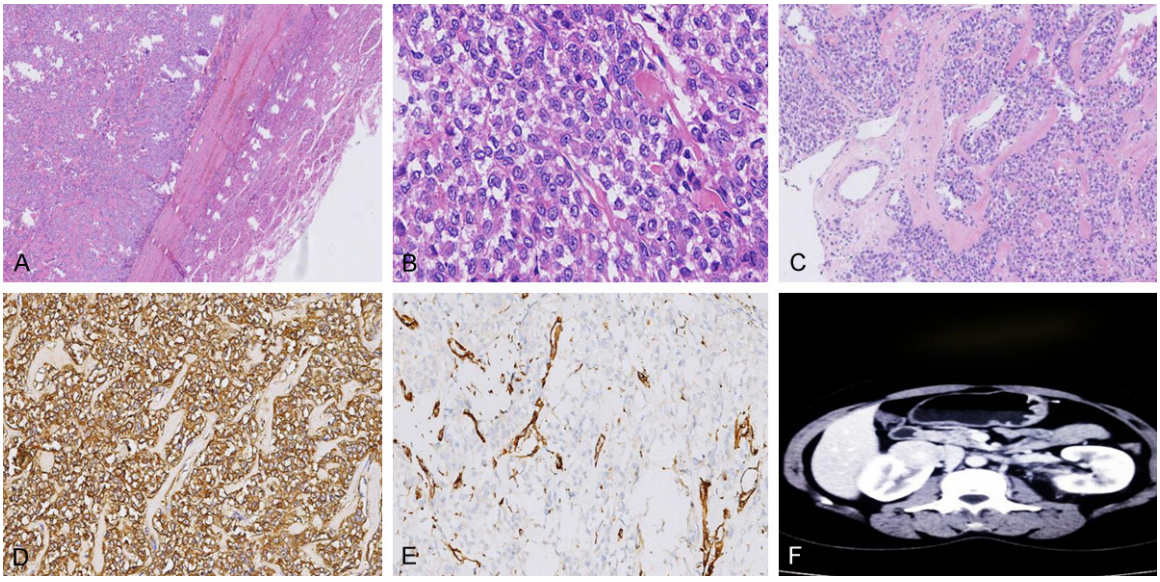

Figure 2. A. The tumor was surrounded by a complete capsule and well-demarcated from the surrounding renal parenchyma HE × 40; B. The tumor mainly composed of round or ovoid cells with a pale or eosinophilic cytoplasm, some of which showed a nest distribution and some showed funicular structure HE × 400; C. Interstitial there are hyaline degeneration of thick-walled blood vessels and collagen fibers HE × 100; D. Immunohistochemical findings: SMA(+) EnVision × 200; E. Immunohistochemical findings: vimentin(-) EnVision × 200; F. Contrast-enhanced CT scan of right kidney.

Microscopic features

The tumor was well-demarcated surrounded by a fibrous psuedocapsule, characterized by diffused distribution of polygonal and round cells, divided by dendritic thin-walled vessels (**Figure 1A**), with an alternation of cellular and paucicellular areas. The tumor cells showed a rich

eosinophilic cytoplasm, nucleus of medium size exhibiting occasional lipid droplets and big bizarre nuclei, nuclear fission (**Figure 1B**). Surrounding the tumor we can see a large area of hemorrhage and necrosis (**Figure 1C**).

Immunohistochemical findings

The neoplastic cells were positive immunostaining for a-inhibin (**Figure 1D**), NSE, S100, vimentin (**Figure 1E**) and CD10. Focal membranous staining was noted for EMA. CD34 outlined the vascular structures. The Ki-67 index was approximately 10%. Negative staining for AE1/AE3, PAX-8, HMB-45, Melan-A, Desmin, P504 and TFE-3.

Pathological diagnosis: Hemangioblastoma.

Gross features

The round neoplasm measured 4.5 × 4 × 3 cm in size, with complete capsule.

Microscopic features

The tumor was surrounded by a complete capsule and well-demarcated from the surrounding renal parenchyma. It was yellowish to red in color and medium to hard in texture (**Figure 2A**). The tumor mainly composed of round or ovoid cells with a pale or eosinophilic cytoplasm, some of which showed a nest distribution and some showed funicular structure (**Figure 2B**). Interstitially, there are hyaline degeneration of thick-walled blood vessels and collagen fibers (**Figure 2C**). Pleomorphic, or bizarre tumor cell nuclei were barely seen, and no nuclear fission was found.

Immunohistochemical findings

The tumor cells showing diffuse immunoreactivity for SMA (**Figure 2D**), Desmin (**Figure 2E**), bcl-2, CD56 and CD99. Focal membranous staining was noted for CD10 and EMA. The Ki-67 index was approximately 3%. CD34 outlined the vascular structures. There was no positive staining for AE1/AE3, EMA, HMB-45, Syn, vimentin, CD10, S-100, WT-1, CD68 and PHH3.

Pathological diagnosis: Epithelioid leiomyoma.

Discussion

The renal primary haemangioblastoma and epithelioid leiomyoma both are very rare. In this case, the patient was diagnosed with hemangioblastoma of the left kidney and epithelioid leiomyoma of the right, since no significant clinical feature indicating the possibility of VHL disease and other systemic disease, we consider the tumors as primary of kidney according to histological characteristics and immunohistochemical results. This is an extremely rare case in clinical and there is no relevant reports in the past. The examination often misdiagnosed the tumor as renal cancer, and the clinical misdiagnosis rate is high [16-20], which is need to be aware of.

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Disclosure of conflict of interest

None.

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References

[1] Plate KH, Vortm Eyer AO, Zagzag D. Von Hippel-Lindau disease and haemangioblastoma Louisdn, Ohgakih, Wiestler OD. WHO classification of tumours of the central nervous system. 4th edition. Lyon: IARC; 2007; 5: 215-217.

[2] Liu Y, Qiu XS, Wang EH. Sporadic the kidney: a rare renal tumor. Diagn Pathol 2012; 7: 49.

[3] Shi H, Li H, Zhen T, Zhang F, Han A. Hemangioblastoma of pelvic cavity: report of a case and review of literature. Int J Clin Exp Pathol 2014; 7: 7054-7058.

[4] Doyle LA, Fletcher CD. Peripheral hemangioblastoma: clinicopathologic characterization in a series of 22 cases. Am J Surg Pathol 2014; 38: 119-127.

[5] Kruizinga RC, van Marion DM, den Dunnen WF, de Groot JC, Hoving EW, Oosting SF, Timmer-Bosscha H, Derks RP, Cornelissen C, van der Luijt RB, Links TP, de Vries EG, Walenkamp AM. Difference in CXCR4 expression between sporadic and VHL-related Sporadic hemangioblastoma. Fam Cancer 2016; 15: 607-16.

[6] Zagoria RJ and Dyer RB. The small renal mass: detection, characterization, and management. Abdom Imaging 1998; 23: 256-265.

[7] Nonaka D, Rodriguez J, Rosai J. Extraneural hemangioblastoma: a report of 5 cases. Am J Surg Pathol 2007; 31: 1545-1551.
[8] Brunocilla E, Pultrone CV, Schiavina R. Renal leiomyoma: Case report and literature review. Can Urol Assoc J 2012; 22: 722-6.
[9] Romero FR, Kohanim S, Lima G, Permpongkosol S, Fine SW, Kavoussi LR. Leiomyomas of the kidney: emphasis on conservative diagnosis and treatment. Urology 2005; 66: 1319.
[10] Jonasch E, McCutcheon IE, Waguespack SG, Wen S, Davis DW, Smith LA, Tannir NM, Gombos DS, Fuller GN, Matin SF. Pilot trial of sunitinib therapy in patients with von Hippel-Lindau disease. Ann Oncol 2011; 22: 2661-2666.
[11] Wei J, Xu Y, Sun L, Sun A. Sporadic hemangioblastoma of kidney: report of a case. Zhonghua Bing Li Xue Za Zhi 2015; 44: 749-50.
[12] Kuroda N, Agatsuma Y, Tamura M, Martinek P, Hes O, alMich M. Sporadic renal Hemangioblastoma with CA9, PAX2 and PAX8 expression: diagnostic pitfall in the differential diagnosis from clear cell renal cell cacinoma. Int J Clin Exp Pathol 2015; 8: 2131-8.
[13] Guo J, Li L, Zhao LH, Li Y, Wang LS, Xiao L, Zhang LY. Clinicopathologic characteristics of sporadic renal hemangioblastoma. Zhonghua Bing Li Xue Za Zhi 2013; 42: 679-82.
[14] Rabadan AT, Hernandez DA, Paz L. Hemangioblastoma located in the posterior incisural space mimicking a tentorial meningioma: a case report. J Med Case Rep 2016; 10: 183.
[15] Kruizinga RC, Van Marion DM, den Dunnenden. Hemangioblastoma of the kidney in a 29-year-old man. Int J Surg Pathol 2012; [Epub ahead of print].
[16] Wei J, Sun L, Sun A, Tao J. Primary leiomyosarcoma of adrenal gland: report of a case. Zhonghua Bing Li Xue Za Zhi 2014; 43: 122-3.
[17] Wei J, Sun A, Tao J, Wang C, Liu F. Primary adrenal leiomyosarcoma: case report and review of the literature. Int J Surg Pathol 2014; 22: 722-6.
[18] Kwon YS, Salmasi A, Han CS and Singer EA. Renal Leiomyosarcoma: Case Report and Review of the Literature. World J Nephrol Urol 2015; 4: 213-217.
[19] Bayar UO, Gun BD, Arikan II. Unilateral ovarian agenesis and clear cell type epithelioid leiomyoma of uterus mimicking ovarian malignancy. J Pak Med Assoc 2014; 64: 79-81.
[20] Sivrioglu AK, Tutar S, Kafadar C, Kara K. The diffusion-weighted imaging of renal leiomyoma. Abdom Radiol (NY) 2016; 41: 1215-6.