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 hemangioblastoma occurs 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.
Microscopic features

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

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 mediumsise 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. Immunohistochemical findings: a-inhibin(+) EnVision × 200; E. Immunohistoch-chemical 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.
Hemangioblastoma complicated with epithelioid leiomyoma in the kidney

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**


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