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CASE REPORT

Clinicopathological and Genetic Study of an Atypical Renal Hemangioblastoma△

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HEMANGIOBLASTOMA (HB), a kind of benign tumor with uncertain histogenesis, is characterized by the presence of stromal cells (SCs) and a rich vascular component.[1] It occurs sporadically, except for about 25% of the cases associated with von Hippel-Lindau (VHL) disease. HB typically occurs in the cerebellum, but it has been reported that HB occasionally occurs in extraneural tissues, such as kidney,[2] adrenal,[3] gastrointestinal tract,[4] soft tissue[5], and so on. We reported an atypical case involving the kidney, which might be misdiagnosed for other renal tumors, especially clear cell renal cell carcinoma (RCC). We also reviewed previously published cases and literature, and made necessary molecular genetic study, in order to investigate its clinicopatholoical features and differential diagnosis, etc.

CASE DESCRIPTION

A 61-year-old male patient was found with a 3.0 cm×2.0 cm sized hypodense mass at the First Affiliated Hospital of Wannan Medical College on May 27, 2015. The tumor was showed in the upper pole of the left kidney on the computed tomography (CT) scan (Fig. 1A, 1B). He had no significant past medical history except for hypertension for several years. The diagnosis of kidney tumor was initially made, but RCC remained to be ruled out. Then partial nephrectomy was performed, which showed, in the upper pole of the left kidney, a solid tumor measured 2.2 cm in diameter with unclear outline and homogeneous section. No other tumors were detected, especially in central nervous system (CNS), and there was no clinical evidence of VHL disease. No tumor recurrence or metastasis occurred within the half-year follow-up period.

Surgically resected renal tumor tissue was fixed in 10% buffered formalin and embedded in paraffin, cut into 5 μm-thick sections with microtome, and stained with hematoxylin and eosin or periodic acid-Schiff stain. SP immunohistochemical staining (IHS) was performed on additional sections for detecting vimentin, AE1/AE3, epithelial membrance antigen (EMA), CK7, CD10, melan-A, HMB45, S100, α-inhibin, neuronspecific endase (NSE), synaptophysin, chromogranin, CD56, CD34, CD31, FⅧ, desmin, smooth muscle actin (SMA), and Ki-67 (Beijing Zhongshan Golden Bridge Biotechnology Co, Ltd). The analyses of VHL gene mutation and hypermethylation

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were performed according to the previously described method.[6]

Macroscopic features demonstrated that part of the left kidney with a size of 3.5 cm×3.2 cm×2.6 cm was cut off. A tumor of 2.2 cm in diameter was found, swelling up the kidney tissue. The cut surface of the tumor was brownish-white, solid and homogeneous (Fig. 1C), with an indefinite outline,

The tumor was ill-demarcated from the surrounding renal parenchyma (Fig. 2A), and in some area the tumor cells broke through the fibrous capsule (Fig. 2B). There was an alternation of cellular and paucicellular areas inside the tumor. The hypercellular areas were full of SCs with pale or eosinophilic cytoplasm (Fig. 2C), with an enriched capillary network enclosed inside (Fig. 2D). The SCs occasionally exhibited lipid droplets, with oval nuclei, small nucleoli and delicate chromatin. Some cells showed pleomorphic, giant, bizarre nuclei, or took a rhabdoid shape (Fig. 2E), but no necrosis or mitoses. The paucicellular areas were composed of enriched capillaries and sparse tumor cells, or completely of fibrous stroma with hemosiderin pigment (Fig. 2F).

The IHS showed that SCs were diffusely positive for vimentin (Fig. 3A), NSE (Fig. 3B), S100 protein (Fig. 3C), α-inhibin (Fig. 3D), and focally positive for CD10 (Fig. 3E), AE1/AE3 and EMA. The tumor cells were strictly negative for CK7, HMB-45, melan-A, chromogranin, synaptophysin, and CD56. The rich and delicate capillary network was outlined well by CD34, CD31, and F Ⅷ. Eosinophilic granula imparted a positive reaction to periodic acid-Schiff stain (Fig. 3F).

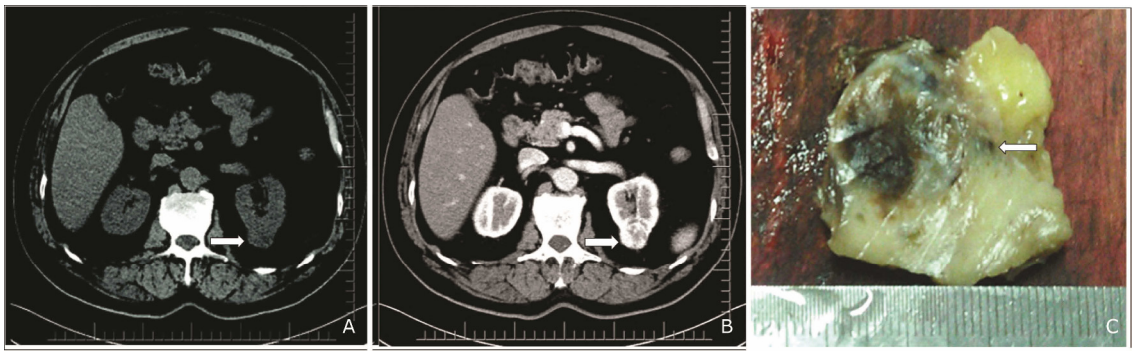

Figure 1. Computed tomography scan and macroscopic findings.
A round hypodense mass in the upper pole of the left kidney (A, B, arrows). The tumor with a diameter of 2.2 cm was ill-demarcated, and its cut surface showed brownish-white in color (C, arrow).

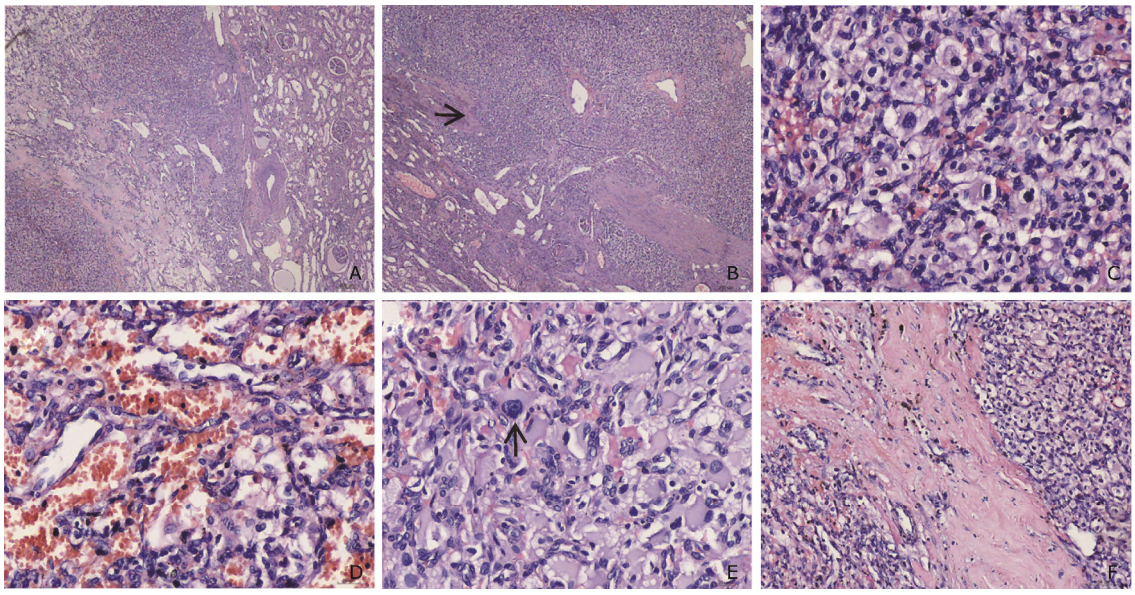

Figure 2. Microscopic findings of HE staining.
The tumor had an indefinite fibrous capsule (A, ×40), and in some area it seemed that tumor cells broke through the capsule (B, ×40, arrow). The tumor consisted of sheets or nests of large polygonal cells with pale or eosinophilic cytoplasm (C, ×400) and abundant arborizing capillary network (D, ×400). Lipoblast-like cells with multiple vacuolization (C), rhabdoid cells (E, ×400, arrow), and hyalinization (F, ×100) were focally seen.