

## Primary Epithelioid Hemangioendothelioma of the Kidney Pelvis

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### 1. Abstract

Epithelioid hemangioendothelioma is a rare type of angiogenic tumor composed of Epithelioid endothelial cells, which is characterized by potentially malignancy and metastasis potential. It is mainly seen in adults, while very rare in children. Tumor can be found in soft tissue or other parts, such as liver and lung, however it is rare in the kidneys. The very report is the first case of that tumor is found in the renal pelvis, which contains only 4 cases of EHE in the kidney. This paper reports one case of EHE in the kidney, and reviews the relevant literatures to discuss its clinical characteristics and differential diagnosis points to better understand the disease.

**2. Keyword:** Epithelioid Hemangioendothelioma; Kidney pelvis

### 3. Case Report

A 30-year-old woman suffered three-month aggravated hematuria with a half-month hospitalization. The urological color doppler indicated a low echo group of left renal pelvis. While, there were no tumor cells were observed by urine shedding cytology and from Ery: +++. Computed Tomography scan, it is found that the partial renal arterial branch is thinner, which is considered as indication of inflammatory lesion (Figure 1A and B). In the left-upper group, the partially widened renal calyx, as well as the internal filling defect, has been found (Figure 1C). Magnetic resonance imaging showed lower perfusion in the middle of the left kidney, which, in combination with CT images, leads to a conclusion of inflammatory lesions (Figure 1D). It is proposed to be used for diagnosing the left renal pelvis mass (3cm\*3cm).

Then partial nephrectomy was performed. Intraoperatively a small, soft, and cystic mass was identified, which was then completely resected and sent for pathologic examination. Intraoperative frozen pathology showed lesions in the left renal pelvis tumor of vascular origin neoplasm. The followings are the proliferation of vascular tissue with a branching network, endothelial cell hyperplasia. With approval from family members, the partial nephrectomy was then performed. The neoplasm pathologic examination showed a macroscopic view: an open solid plane; gray, red, medium texture,

partial surface filth. From the light microscopy scan, the surface of the lesion was found to be more hemorrhagic necrosis, and the subsurface of the vascular tissue with hyperplasia was branched; in addition to these, the hyperactive endothelial cells as well as mucinous and collagen-like blood vessels were found. The diagnosis is the changes of vascular source in left renal pelvis, which is considered as epithelioid hemangioendothelioma. Immunohistochemistry: CD10: (-) ; CD31(+); CD34(-) ; CK(-) ;D2-40(-) ; ERG(+); ki-67(+20%). Two weeks after surgery, the hematuria disappeared and the ischemia in the left kidney was improved from CT scan and the ery is negative.



Figure1A

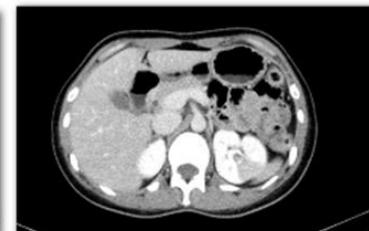


Figure1B



Figure 1C

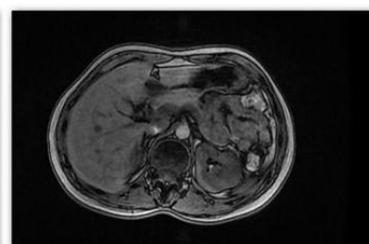


Figure 1D

**Figure 1:** Then partial nephrectomy was performed. Intraoperatively, a small, soft, and cystic mass was identified, which was then completely resected and sent for pathologic examination. Intraoperative frozen pathology showed lesions in the left renal pelvis tumor of vascular origin neoplasm.

### 4. Discussion

Epithelioidhemangioendothelioma, a rare potentially malignant tumor that first named in 1982, could occur in many parts of the

body, of which the main parts are soft tissue and liver[1]. EHE is a rare blood vessel tumor that contributes to less than 1% of all hemangiomas, and the incidence of EHE is less than one in a million, which has epithelioid tissue cell appearance that originated from vascular endothelium or anterior endothelial cells [2][3]. Renal pelvis epithelioid hemangioendothelioma is extremely rare in clinic, and this case is exactly the first one in the world. As pediatric cases reported in 2013, similar symptoms of tumor near the renal pelvis was found, which led to hematuria [4]. Two of the remaining three patients were asymptomatic, and medical examination showed that, the other one presented with abdominal pain and no special symptoms [5][6]. Thus, EHE disease is hidden, and the specific performance of which is deficient. This case was the first case in which the patient had the tumor was exactly in the renal pelvis (Figure 2). Immunohistochemical stains were positive for vascular endothelial markers indicated that multiple endothelial proteins may help identify EHE. CD31 and CD34 showed better sensitivity and specificity than endothelial marker. It was reported that CD34 was expressed in more than 90% of vascular tumors, which indicates that this marker has poor specificity. By contrast, CD31 is considered as a relatively specific marker of vascular tumor. Fli-1 combined with CD31 has 100% sensitivity and 92% specificity to EHE, making it an ideal choice for diagnosis of EHE [7].

In term of treatment of this case, the small volume of tumor was covered with a membrane, and the partial nephrectomy was reserved. The tumor size and location seem to be the only considerable factors to decide the surgical approach (Figure 3). The effect of chemotherapy and radiation therapy has not been completely confirmed in the kidneys of EHE. The studies have shown that sunitinib is effective in some degree histological examinations revealed mainly epithelioid cells [8]. It has been reported that bevacizumab combined with nanoparticle albumin and paclitaxel has good tolerance and stability in the treatment of invasive metastatic p-ehe, and anti-angiogenesis therapy is an effective method for the treatment of EHE [9].

The world health organization (WHO 2002) defines EHE as a locally invasive tumor with metastatic potential [10] A study of the prognosis of 49 patients with ehe showed that the ehe could be classified as low risk and high risk based on mitotic activity of tumor cells and tumor size, and those tumors with >3 mitotic figures/50 high power fields and size >3.0cm had the worst prognosis [11]. The relative mortality of epithelioid hemangioendothelioma varies with the location. The mortality rate of soft tissue location was 13%, while, the death rate of lung and liver disease was 65% and 35%, respectively [12]. Studies have shown that malignant progression of ehe is associated with changes in the expression of TP53, MDM2, cav1 and VEGF [13]. Survival data of kidney lesion survival data is not yet clear, because there are very few documented cases; and more information on the prognosis of renal pelvis disease

prognosis is unknown. However in this case, endothelial markers ERG is worth mentioning, since they are a family of transcription factors. And ki-67 is a 20% proliferation index, which leads to the considering the tumor as potentially malignant.

Although the results of imaging and urinalysis 2 months after surgery in this patient confirmed the effectiveness of surgical resection, long-term follow-up was still required.

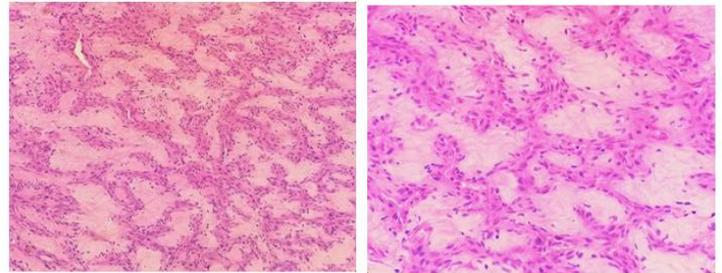


Figure 2A (HE x200)

Figure 2B (HE x400)

**Figure 2:** The histological characteristics of epithelioid vascular endothelioma are the proliferation of ovoid endothelial cells, which are distributed in the mucous clear matrix. The tumor cells are proliferating vigorously, with abundant cytoplasm and vacuoles. Nuclear hyperchromatism, no atypia and mitotic activity were found. Erythrocytes were occasionally seen in the lumen.

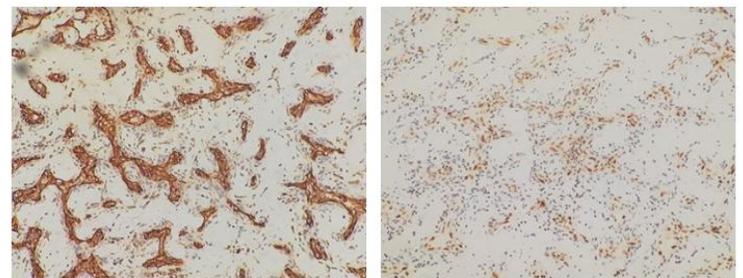


Figure 3A (CD31 x400)

Figure 3B (ERG x400)

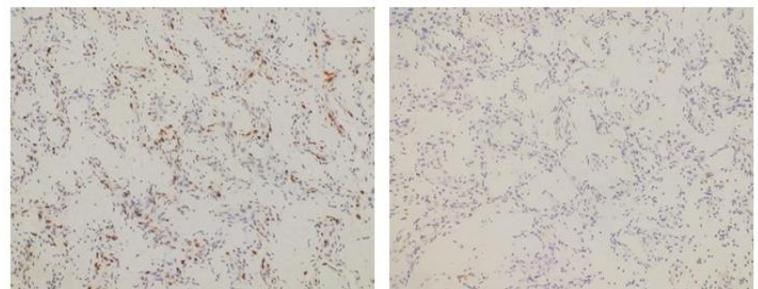


Figure 3C (ki-67 x400)

Figure 3D (CD10 x400)

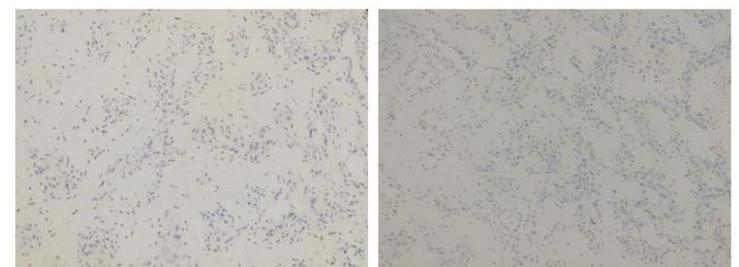


Figure 3E (CD34 x400)

Figure 3F (CK x400)

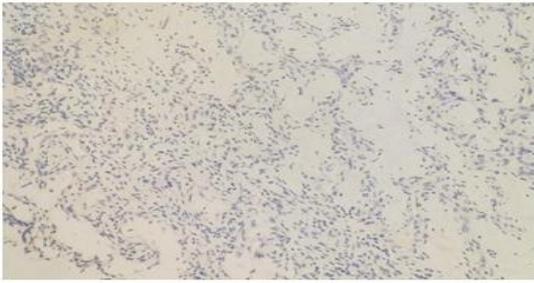


Figure 3G (D2-40 x400)

**Figure 3:** Immunohistochemistry of epithelioid hemangioendothelioma

## 5. Conclusion

EHE is a rare tumor, especially in the renal pelvis. The first renal pelvis EHE was described in this paper. The patient of which were found to be with only one clinical manifestation of hematuria. Due to specific clinical features, imaging findings and tumor location, the surgical resection was then performed. For the very patient, although EHE may be histologically an intermediate malignant, long-term follow-up is clinically important because of the potential malignancy of the tumor.

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