Anastomosing Hemangioma Mimicking Renal Cell Carcinoma

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Anastomosing hemangioma (AH), a rare benign vascular tumor, is a newly recognized variant of capillary hemangioma. In the microscopic examination, this tumor has characteristic feature of the unique anastomosing sinusoidal capillary sized vessels. It can be misdiagnosed as a malignancy such as renal cell carcinoma or angiosarcoma. Herein, we report a case of AH originating in the right kidney of a 43-year-old man, which was initially considered as cystic renal cell carcinoma on computed tomography (CT). The patient underwent laparoscopic radical nephrectomy, but pathologic result was AH of the kidney. There was no evidence of recurrence or metastasis 5 months after the surgery. (Korean J Urol Oncol 2016;14:88-92)

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In 2009, Montgomery and Epstein reported 6 cases of new variants of benign vascular tumors involving the kidneys, perinephric adipose tissue and testes, which the authors designated as anastomosing hemangioma (AH).¹ Subsequently, several cases of this novel vascular tumor have been reported occurring not only in the genitourinary tract but also sporadically in the ovary, adrenal gland, liver and gastrointestinal tract.²⁻⁴ Microscopically, this tumor is composed of anastomosing sinusoidal capillary sized vessels with scattered hobnail endothelial cells within a framework of non-endothelial supporting cells. AH in the kidney is a very rarely recognized benign neoplasm. But unfortunately, renal AH is not easily diagnosed preoperatively because of its general small size and nonspecific imaging findings.⁵ Renal AH commonly develops in the context of chronic kidney disease (CKD) and shares both the clinical presentation and imaging findings with a malignant tumor such as renal cell carcinoma or angiosarcoma. Herein, we would report an anastomosing hemangioma mimicking renal cell carcinoma in middle-aged diabetic man with CKD.

CASE REPORT

A 43-year-old man was consulted to department of our urology for an incidentally detected mass in the right kidney on computed tomography (CT), which was taken during orthopedic treatment of Charcot foot caused by complication of diabetes. Urinalysis and urine culture were nonspecific. Routine blood examination revealed normal range of white blood cell count (4800 cells/μl) and decreased hemoglobin level (9.3g/dl). Preoperative blood chemistries were unremarkable except for an elevated serum creatinine (1.81mg/dl), and decreased estimated glomerular filtration rate (eGFR) (44.8ml/min/1.7m²). Pre-enhanced CT image showed a 4.3x3.8cm, well circumscribed mass on the peri-hilar portion of the right kidney and small calcifications on pancreatic head portion (Fig. 1A). Contrast-enhanced CT showed a well demarcated mass with obviously in-
Fig. 1. (A) Pre-enhanced CT image shows a 4.3x3.8cm, well circumscribed mass on the peri-hilar portion of the right kidney (yellow arrow) and small calcifications on pancreatic head portion (red arrow). (B) Contrast-enhanced CT shows a well-defined mass with obviously annular and nodular enhancement which is suggestive of renal cell carcinoma (yellow arrow). (C) Delayed CT image shows a right renal mass with homogeneously persistent enhancement and a well-circumscribed boundary (yellow arrow).

Fig. 2. Histological evaluation revealed that (A) Macroscopic image shows the 4.3x2.7cm tumor in the right renal hilum with well-demarcated margins and a spongy consistency (white arrows). (B) At low power (H&E x40), the lesion is well demarcated from normal renal tissue and a loosely lobulated architecture. (C) H&E x100 image shows an anastomosing vascular pattern composed of sinusoidal capillary sized vessels. (D) At higher magnification (H&E x200), the cellular zones were composed of anastomosing sinusoidal-like irregular vessels lined by hobnail endothelial cells (arrow) and a flat endothelium focally, seated in a framework of fibrous supporting stromal tissue.
tense annular and nodular enhancement (Fig. 1B). On delayed CT image, the mass showed homogeneously persistent enhancement and a well-circumscribed boundary (Fig. 1C). The CT findings were suggestive of renal cell carcinoma. We performed laparoscopic radical nephrectomy after detailed discussion because the tumor was too close to the main renal vein to do partial nephrectomy. Grossly, cut section of the kidney showed 4.3x2.7cm tumor with well-demarcated margins and a spongy consistency in the right renal hilum (Fig. 2A). Microscopically, the tumor was well demarcated from normal renal tissue but had no pseudocapsule. At low power field, the lesion demonstrated a loosely lobulated architecture (Fig. 2B). At higher magnification field, the tumor had a “sieve-like” anastomosing vascular pattern composed of sinusoidal capillary sized vessels (Fig. 2C), and the vessels were lined by hobnail endothelial cells and a flat endothelium focally (Fig. 2D). Cytologically, the tumor cells were overall bland with only a slight degree nuclear enlargement but lacking both marked nuclear atypia and any mitotic figures. Immuno-histochemical studies showed that the tumor cells were diffusely positive for CD31, CD34, factor VIII-related antigen, but negative for HMB45, Cytokeratin, EMA and D2-40 (Fig. 3). The overall histopathological examination confirmed AH of the right kidney. The patient discharged home 14 days after the surgery due to further orthopedic care, and there was no evidence of recurrence or metastasis 5 months after the surgery.

**DISCUSSION**

In general, hemangioma is a relatively common mesenchymal
tumor that typically occur skin, subcutaneous soft tissue, and occasionally in viscera. The liver is the most frequent involvement site among the visceral organs. However, primary benign vascular tumors of the kidney are uncommon. Most of the reported renal hemangiomas have been histologically classified as capillary or cavernous subtype.\textsuperscript{6} AH is a morphological variant of capillary hemangoma firstly described by Montgomery and Epstein. The term “AH” described the histological observation of anastomosing pattern of capillary sized vascular channels reminiscent of splenic sinusoids.\textsuperscript{1}

A total of 25 cases of this peculiar vascular tumor of the kidney have been reported in the English-language literature since the first report in 2009.\textsuperscript{7,8} Average age of presentation is 53 years old and the lesions are mostly unilateral, isolated with only one bilaterally. These tumors in the kidney show a slight male predominance with a male to female ratio of 1.4:1. Median tumor size of the previously reported 25 cases was 2.0cm (ranges from 0.6-5.0cm). The clinical presentation is nonspecific, which includes intermittent hematuria, abdominal pain, and lower urinary tract symptoms. Our patient also had no symptoms. A majority of this lesion is detected incidentally, during radiographic investigations for other unrelated reasons. The current case was also detected incidentally at the imaging study which was taken prior to the orthopedic surgery.

The association between acquired renal cystic disease secondary to dialysis and malignant renal epithelial neoplasm is well documented. There was a wide spectrum in the cause of the CKD in the cases reported which precludes the possibility of a specific underlying cause.\textsuperscript{9,10} Remarkably, eight (30.8\%) of reported cases have preexisting CKD, which is suspected to a tendency of AH to develop in CKD. Our patient also had CKD caused by poorly controlled diabetes. Initially, his serum creatinine level and eGFR were 1.81mg/dl and 44.8ml/min/1.7m\textsuperscript{2}, respectively. To date, the exact pathogenesis of CKD-associated AH arising from a specific underlying cause is unknown. What is known is the propensity of kidneys damaged by CKD to develop not only renal cell carcinomas but also benign mesenchymal neoplasms. Therefore, AH is not unique to CKD as observed in our case.

Imaging information about AH in the kidney is limited. Contrast-enhanced CT imaging cannot reliably differentiate renal AH from other aggressive renal neoplasms such as angiosarcoma and renal cell carcinoma. Consequently, AH is often underdiagnosed prior to surgery and usually treated with nephrectomy.\textsuperscript{11,12} Tao et al reported a case of AH with detailed unenhanced CT and contrast enhanced CT scan image information.\textsuperscript{13} In this literature, the unenhanced axial CT scan showed a left renal sinus mass with a round, well-circumscribed figure that appeared to be heterogeneous. In the arterial phase of the contrast-enhanced CT scan, the boundary of the mass showed a strong annular and nodular enhancement. In the venous phase, the mass demonstrated further intense enhancement. On delayed images, the lesion showed homogeneously persistent enhancement and a well-circumscribed boundary. When we reviewed our case, we could find very similar CT findings to their report.

Macroscopically, AH is usually well demarcated but non-encapsulated with a mahogany brown spongy consistency and is typically located in the hilum. There is no grossly evident necrosis or vascular invasion. Microscopically, most lesions are well circumscribed from normal renal tissue. At low power, the lesion demonstrates a loosely lobulated architecture, with alternating cellular zones and hypocellular areas. The cellular areas comprise proliferations of capillary sized vessels in an anastomosing pattern, while the hypocellular areas comprise loose stromal tissue with elastic thin-walled blood vessels. At higher magnification, the vessels are lined by hobnail endothelial cells and a flat endothelium focally. Zones of sclerosis and deposition of collagen between the sinusoidal vessels are observed. Cytologically, a slight degree of nuclear enlargement is found, but cellular atypia, multi-layering of endothelial cells and apoptotic figures or mitotic activity are not detected. Immuno-histochemical stains show that the tumor cells are diffusely positive with endothelial markers CD34 and CD 31, and factor VIII-related protein, the stroma cells are positive for SMA, and Ki-67 showed low proliferation activity of the tumor cells.\textsuperscript{12,14}

All the reported cases were treated with surgery and this tumor showed excellent prognosis after surgical resection.\textsuperscript{11} The follow up of reported cases ranged from 3 to 156 months (average: 26.1 months) and none have experienced tumor recurrences, metastases, or death due to the renal tumor. So AH in the kidney seems to be a biologically benign tumor, however because most of these cases are recently recognized, long term surveillance of more cases is warranted to define a definitive conclusion on its biological behavior.
REFERENCES


