CASE REPORTS

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Anastomosing hemangioma of the kidney: a case report

Anastomozirajući hemangiom bubrega

Aleksandar Lazić*, Predrag Marić*, Jelena Tadić†, Božidar Kovačević‡§, Bojana Jovanović‡, Bojana Krća‖, Mirjana Mijušković†§

Military Medical Academy, *Clinic for Urology, †Clinic for Nephrology, ‡Pathology and Forensic Medicine Institute, ‖Institute of Radiology, Belgrade, Serbia; §University of Defence, Faculty of Medicine of the Military Medical Academy, Belgrade, Serbia

Abstract

Introduction. Renal anastomosing hemangioma (AH) is a very rare vascular tumor. This type of tumor is named so because of its unique histological structure, which resembles splenic sinusoidal structures. It may mimic malignant neoplasms, like angiosarcoma, which is why clinical and radiological examinations are insufficient for accurate diagnosis. **Case report.** We present a case of a 39-year-old male admitted to the Clinic for Nephrology at the Military Medical Academy, Belgrade, Serbia, for considering a living-related kidney transplant due to the development of end-stage renal disease (ESRD). His father was identified as the prospective donor. During the patient's pre-transplant evaluation, a suspect tumorous lesion in the left kidney was observed, and multislice computed tomography scanning of the abdomen and small pelvis was performed. This imaging revealed a lobulated soft tissue lesion measuring approximately 25 × 15 mm in the lower pole of the left kidney. Based on this finding, it was decided to proceed with a left nephrectomy to obtain a histopathological assessment of the lesion. The histopathological examination, complemented by immunophenotyping, led to the diagnosis of an AH in the kidney. **Conclusion.** The presented case emphasizes the importance of urgent surgical resection of the tumor mass in order to confirm the diagnosis and avoid any delay in patients scheduled for kidney transplantation.

Key words:
diagnosis, differential; hemangioma; immunohistochemistry; kidney failure, chronic; kidney transplantation; multidetector computed tomography; vascular neoplasms.

Apstrakt

Uvod. Anastomozirajući hemangiom (AH) bubrega je veoma redak vaskularni tumor. Ovaj tip tumora je tako nazvan zbog njegove jedinstvene histološke strukture, koja podseća na sinusoidne strukture slezine. Može ličiti na maligne neoplazme, kao što je angiosarkom, zbog čega kliničko i radiološko ispitivanje nije dovoljno za tačnu dijagnozu. **Prikaz bolesnika.** Prikazujemo 39-godišnjeg muškarca, koji je primljen na Kliniku za nefrologiju Vojnomedicinske akademije u Beogradu, Srbija, radi razmatranja transplantacije bubrega od živog srodnika zbog razvoja terminalne bubrežne bolesti. Kao potencijalni donor bubrega određen je otac bolesnika. Tokom pretransplantacijske procene bolesnika, primećena je sumnjiva tumorska lezija u levom bubregu i urađena je multislajsna kompjuterizovana tomografija trbuha i male karlice. Ovim pregledom je u donjem polu levog bubrega otkrivena lobulirana mekotkivna promena, veličine približno 25 × 15 mm. Zbog navedenog nalaza, odlučeno je da se uradi uklanjanje levog bubrega, radi histopatološke potvrde promene. Patohistološkom analizom, dopunjenom imunofenotipizacijom, dijagnostikovan je AH u bubregu. **Zaključak.** Prikazani slučaj naglašava značaj hitne hirurške resekcije tumorske mase radi potvrde dijagnoze i izbegavanja odlaganja planirane intervencije kod bolesnika kod kojih je planirana transplantacija bubrega.

Ključne reči:
dijagnoza, diferencijalna; hemangiom; imunohistohemija; bubreg, hronična insuficijencija; transplantacija bubrega; tomografija, kompjuterizovana, multidetektorska; krvni sudovi, neoplazme.

Introduction

Primary vascular tumors of the kidney are exceedingly rare, which represents a paradox considering the kidney's robust blood supply, as it is believed to receive nearly one-quarter of the cardiac output. The majority of such cases have been documented as isolated reports in medical literature. Montgomery and Epstein [1] have described a new

Correspondence to: Aleksandar Lazić, Military Medical Academy, Clinic for Urology, Crnotravska 17, 11 000 Belgrade, Serbia. E-mail: medalazic@gmail.com

variant of renal capillary hemangiomas, characterized by distinctive features that combine sinusoidal and hobnail patterns, typical of hemangiomas of the skin and soft tissues. Hemangiomas, more prevalently found in the skin and subcutaneous tissues, are less frequent in visceral locations, with the liver being the most common site [2]. Histologically, hemangiomas are broadly categorized into cavernous and capillary types [3, 4]. A novel variant of renal capillary hemangiomas, characterized by distinct features amalgamating both sinusoidal and hobnail patterns, typically observed in skin and soft tissue hemangiomas, was delineated by Montgomery and Epstein [1] in 2009. This tumor type was designated as "anastomosing hemangioma" (AH) due to its unique histological structure reminiscent of splenic sinusoids. AH was acknowledged in the 2020 World Health Organization classification of soft tissue tumors. Isolated renal AH accounts for 22% of cases and is typically identified as an incidental finding during imaging. The lesion usually presents as solitary, although instances of bilateral and multifocal occurrence have been documented. The radiological characteristics of AH are nonspecific, which complicates the presurgical diagnostic process. Differential diagnoses, which depend on the location of the AH, include, among others, angiosarcoma, clear-cell renal cell carcinoma (RCC), ectopic paraganglioma, and pheochromocytoma [5].

Interestingly, AHs are predominantly observed in patients with end-stage renal disease (ESRD) [6]. Initially believed to be exclusive to the genitourinary system, with a particular affinity for the kidneys [1, 7, 8], AH has since been identified in other parts of the body, including the testes, thighs, the abdominal wall [1], ovaries [7], the adrenal gland [8], the liver, and the gastrointestinal tract [9]. Despite its benign nature, the radiological profiles of AHs closely resemble those of RCCs [1, 10], the most prevalent form of kidney cancer in patients with ESRD [11].

We present a case of AH, definitively diagnosed by histological examination after nephrectomy.

Case report

A 39-year-old male was admitted to the Clinic for Nephrology at the Military Medical Academy, Belgrade, Serbia, for a potential living-related kidney transplantation (KT) from his father as the donor. The patient has been on maintenance hemodialysis since October 2022. Primary kidney disease was focal segmental glomerulosclerosis.

During the pre-transplantation evaluation, a suspicion of a tumorous change in the left kidney led to a multislice computed tomography scan of the abdomen. The scan revealed a lobulated soft tissue change of approximately 25 × 15 mm in the lower pole of the left kidney, which showed a post-contrast peripheral increase in density. Additionally, a similar lesion of about 10 mm was identified in the lower pole of the right kidney, raising the possibility of primary and infiltrative lesions (Figure 1).

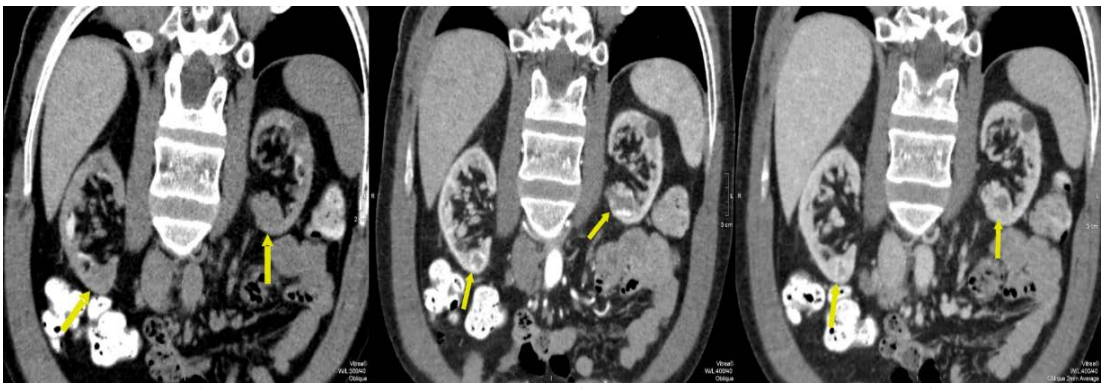
A histopathological examination, complemented by immunophenotyping, was performed.

Macroscopically, the kidney, with a fibrous and fatty capsule and a segment of the ureter measuring 60 mm in length, was observed. On sectioning the renal tissue near one pole, a clearly demarcated, hemorrhagic-looking field measuring 10 × 15 mm was present, processed entirely in molds marked with numbers 3–5.

Microscopically, the renal tissue contained a benign mesenchymal tumor of the hemangioma type, with the largest diameter of up to 15 mm. The tumor was mostly well-demarcated and partly expansively growing with an irregular border towards the surrounding hilus and renal tissue. Histologically, the tumor comprised anastomosing channels of vascular spaces of varying widths with thin walls lined by flat or low cuboidal endothelium. The central zone of the tumor showed sclerosis with foci of hemorrhage and/or hemorrhagic necrosis (Figure 2).

Immunohistochemical analyses yielded diffuse positivity for CD31, CD34, factor VIII, and vimentin in the tumor cells, while reactions for pan-cytokeratin (PanCK), CD10, CK7, α-methylacyl-CoA racemase (AMACR), RCC, and epithelial membrane antigen (EMA) were negative. The proliferative potential of the tumor cells, determined using antibodies for Ki67, was less than 2% (Figure 3).

The histopathologic appearance, along with the immunophenotypic feature of this tumor, indicated a diagnosis of AH of the kidney.



A) **B)** **C)**

Fig. 1 – A) In the lower pole of both kidneys, lobulated soft tissue hypodense lesions are visible, measuring 25 × 15 mm on the left and up to 10 mm on the right. The described changes exhibit pronounced marginal nodular post-contrast enhancement in density during the arterial phase of the examination (B), with slight washout in the venous phase of the examination (C).

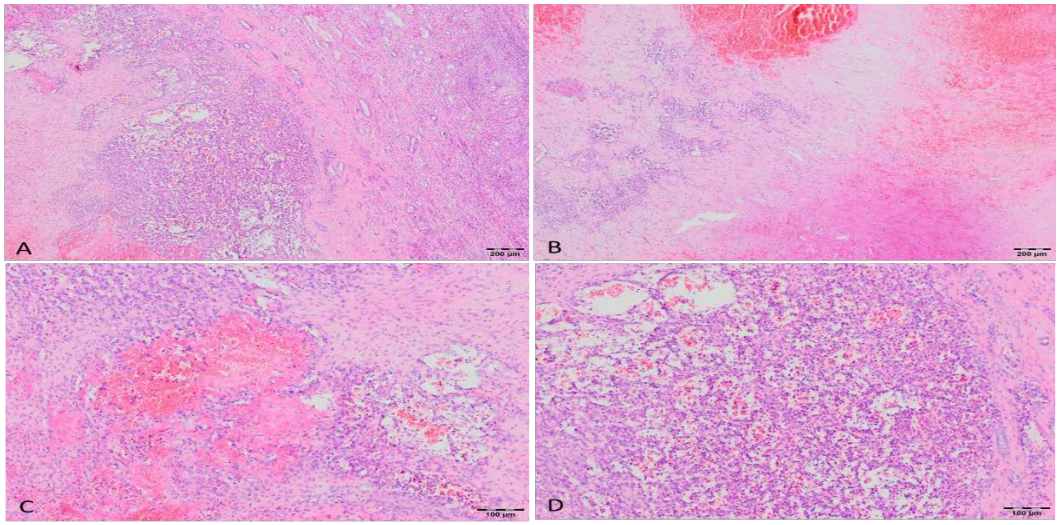


Fig. 2 – A) Expansive growth of anastomosing hemangioma with irregular border to the renal parenchyma [hematoxylin and eosin (HE) staining, ×4]; B) and C) sclerosing zone with foci of hemorrhage (HE staining, ×4 and ×10, respectively); D) focus of prominent thin-walled anastomosing vascular structures, lined by single layer of bland endothelial cells (HE staining, ×10).

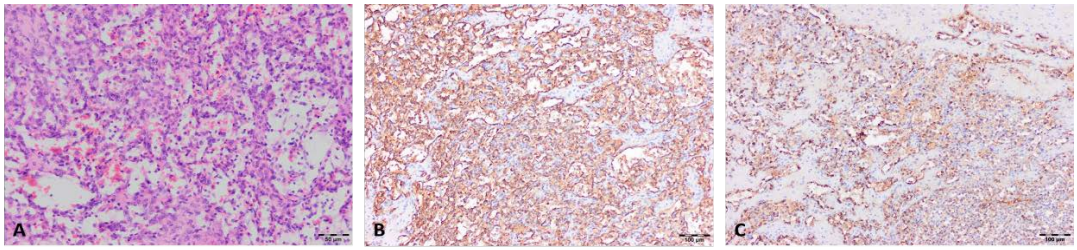


Fig. 3 – A) Higher magnification of prominent thin-walled anastomosing vascular structures, lined by single layer of bland endothelial cells [hematoxylin and eosin (HE) staining, ×20]; B) endothelial cells lining thin-walled anastomosing vascular channels positive to CD34 marker (HE staining, ×10), and C) factor VIII (HE staining, ×10).

Given these findings, due to the differential diagnostic suspicion of RCC and in consultation with a urologist, a decision was made to perform a left nephrectomy for histopathological verification of the change in the lower pole. Given that the patient was already on maintenance hemodialysis and was anuric, there was no need for a spare operation on the left kidney.

The postoperative course was marked by transient fevers, peaking at 38 °C, and the patient was initially treated with third-generation cephalosporins (ceftazidime) and transiently with carbapenems. The blood culture sample showed no increase in bacterial colonies until discharge. The drain from the surgical wound was removed on the second postoperative day, and the wound healed primarily. A secondary anemic syndrome was corrected with the administration of erythropoietin. Four months after the surgical treatment, the patient underwent a successful KT in February 2024.

Discussion

Despite the kidney's good vascularization, primary vascular tumors of the kidney are rare. AHs predominantly affect middle-aged adults, with a mean age of 52.6 years, ranging from 21 to 83 years, and are most commonly observed in individuals in their fifth to sixth decades. Literature indicates a slight male predominance in renal hemangioma cases, with a male-to-female ratio of 1.8 : 1 [12]. The presented case aligns with this demographic, involving a 39-year-old male.

Lesions in this context are typically unilateral, though one bilateral instance has been documented [7]. These tumors can affect both kidneys equally and may involve any renal region. Recent studies have identified activating mutations of *GNA* genes that drive the molecular pathogenesis of AHs [13].

The clinical manifestations of these tumors are nonspecific. While hemangiomas of the bladder have been associated with conditions such as tuberous sclerosis, Klippel-Trenaunay syndrome, and Sturge-Weber syndrome [14], there is no established correlation between AHs of the kidney and any systemic or syndromic conditions [3, 15]. Common clinical presentations include intermittent hematuria, flank pain [1], and lower urinary tract symptoms [4]. However, many lesions are incidentally detected during radiographic investigations for unrelated reasons [7] or due to other diseases at the time of nephrectomy [3, 7]. Nevertheless,

specific features can assist the radiologist in forming a diagnosis. A solid renal mass exhibiting significant T2 hyperintensity, almost resembling a cyst, may serve as an indicator [16]. In the presented case, the patient did not exhibit any symptoms associated with the neoplastic change in the kidney, which can be explained by the fact that the patient was anuric and that the tumor was relatively small in size. Radiological diagnostic procedures prompted suspicion of renal lesions, with nephrectomy leading to a definitive diagnosis. To the best of our knowledge, this is the second case of AH during a medical workup before KT [17].

Approximately 37% of AH cases reported in the literature are associated with ESRD [3, 7, 15]. Büttner et al. [18] found eight hemangiomas, all histologically capillary type with a sinusoidal pattern, indicative of AH in a retrospective examination of 90 nephrectomy specimens from ESRD patients. This, coupled with our case, suggests a propensity for these particular hemangiomas to develop in ESRD. The pathogenesis of ESRD-associated AH remains unclear, but there is a known tendency for kidneys damaged by chronic disease to develop both epithelial renal tumors and benign mesenchymal tumors [18]. It is also important to note that AH can develop in kidneys without chronic disease.

Macroscopically, AHs typically appear well-demarcated yet non-encapsulated with a mahogany brown, spongy consistency, usually situated in the renal hilum [1, 3]. Microscopically, they are well-marginated, often with normal renal tubules entrapped at their periphery [3, 7]. Cytologically, the tumor cells typically lack malignant features, and immunochemically, they consistently express endothelial markers while being negative for other markers, confirming their endothelial origin [1, 4]. These characteristics were also validated in the presented case, with positivity for CD31, CD34, factor VIII, and vimentin in the tumor cells, while reactions for PanCK, CD10, CK7, AMACR, RCC, and EMA were negative.

Differential diagnosis includes malignant vascular tumors such as angiosarcoma and Kaposi sarcoma. Both of these entities, which are infrequent in renal presentations, may exhibit hyaline globules. Specifically, the former often demonstrates an anastomosing vascular pattern and hobnail endothelial cells. This particular histological presentation can lead to diagnostic confusion with AH, particularly when examining needle core biopsy specimens. AH can also be confused with other vascularized renal neoplasms like clear cell RCC [7], hemangioblastoma [19], and glomus tumor [20].

The median size of renal AHs is reported to be 15 mm, with most lesions being smaller than 40 mm [13]. According to this criterion, our case aligns with those previously described in the literature, given that the size of the tumor lesion measured 25 mm in its largest diameter.

Current treatment guidelines for renal hemangioma consider tumor size, location, and patient symptoms [2]. Although AH is benign, surgical intervention is recommended due to its similarities with malignant lesions. In our case, we adhered to these guidelines, opting for left radical nephrectomy. Given the patient's diminished renal function and the need for adequate surgical margins, radical nephrectomy was deemed necessary despite the tumor's small size. It was decided to monitor the lesion in the patient's right kidney, as it shares the same radiological characteristics as the lesion in the operated kidney. We feel disposed to emphasize the importance of conducting regular ultrasound monitoring and, if necessary, computed tomography scanning of the right kidney lesion.

Conclusion

This specific subtype of hemangioma is likely more prevalent than initially perceived and frequently arises in the setting of end-stage renal disease. However, the morphological similarities between anastomosing hemangiomas and renal neoplasms like renal cell carcinomas and angiosarcoma may present a significant challenge. The presented case underlines the importance of prompt surgical resection of suspicious malignant tumor changes in the kidney in order to confirm the diagnosis and avoid any delay in patients scheduled for kidney transplantation. The presented patient underwent a successful kidney transplantation in February 2024, four months after a left-sided nephrectomy.

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