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Juxtaglomerular Cell Tumor as a Rare Cause of Malignant Hypertension Surgically Curable By Robot-Assisted Partial Nephrectomy: A Case Report

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Abstract

A tumor originating from juxtaglomerular cells can produce renin, thus also called Reninoma, which leads to hypertension by secondary hyperaldosteronism. Here, we report the case of a 27-year-old female presenting with a 32mmx29mmx27mm centrally located, solid kidney tumor in the middle pole of the left kidney. The tumor was clinically suspicious for an oncocytoma with the differential diagnosis of a renal cell carcinoma. The patient suffered from a previously known persistent arterial hypertension for over six years, despite a combination therapy with three antihypertensive drugs. After robot-assisted partial kidney resection, the hypertension resolved without antihypertensive medication. Histopathologically, a juxtaglomerular cell tumor, reninoma, was diagnosed. Since 1967, less than one hundred clinical cases of juxtaglomerular cell tumors associated with hypertension have been reported. Previous case reports mainly describe total nephrectomy as the curative therapy of choice, whereas nowadays kidney-sparing procedures should be the preferred treatment. Thus, we conclude that robot-assisted partial kidney resection is a minimally invasive, surgically safe and curative treatment option for hypertension caused by juxtaglomerular cell tumors.

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Introduction

Reninoma is a tumor of juxtaglomerular cells which produces renin, leading to renin-dependent hypertension by secondary hyperaldosteronism. The overexpression of renin is caused by the dysfunction of juxtaglomerular cells. Hypertension caused by a reninoma was first described by Robertson et al. in 1967 [1-8]. Since 1967, less than one hundred clinical cases of juxtaglomerular cell tumors associated with hypertension have been reported. The average age at diagnosis is 27 years. Epidemiologically, juxtaglomerular cell tumors are more common in females with a gender ratio of 2:1. The diagnosis is typically preceded by hypertension which is not responsive to antihypertensive medications. Histopathological examination is the gold standard for diagnosing juxtaglomerular cell tumor, as other renal tumors may also raise plasma renin levels, such as renal cell carcinoma and Wilms tumor. Despite causing severe hypertension, juxtaglomerular cell tumors are generally considered benign tumors [4,5,10]. Rare cases of metastasis, recurrence and capsular or vascular invasion are known [1,2,4,10,12].

Case Report

A 27-year-old female patient with a body-mass-index of 24 presented with persistent arterial hypertension and blood pressure values of 200/140 mmHg for over 6 years. The combination of Valsartan, Amlodipine, and Spironolactone was initiated at the time of diagnosis. Despite this treatment, the patient's arterial hypertension persisted so far. Ultrasonography showed at initial diagnosis of hypertension neither an evidence of renal artery stenosis nor an evidence of a kidney tumor. In the recent reevaluation, MRI revealed the presence of a 32mmx29mmx27mm centrally located, solid kidney tumor in the middle pole of the left kidney, not constricting the renal artery (Figure 1). Radiologically, the mass was suspicious for oncocytoma, with a differential diagnosis of renal cell carcinoma. The blood tests showed hyperaldosteronism and highly elevated plasma renin but no electrolyte imbalance and normal potassium values. Informed consent was obtained including a discussion of hypertension associated with the kidney tumor. A robot-assisted partial kidney resection was performed without complications and a warm ischemia time of 11 minutes. Postoperatively, regular sonographic follow-ups of the left kidney were performed, and antihypertensive therapy was discontinued. Even though antihypertensive medication was omitted immediately after surgery, the patient was discharged with normotonic blood pressure values of 120/80 mmHg. After discharge, there was no further need for antihypertensive medication. Histopathologically, a juxtaglomerular cell tumor or reninoma was diagnosed. The tumor presented uniformly round to polyclonal, slightly eosinophilic cells with glomoid appearances, numerous focally sclerosed branching blood vessels, pronounced lymphocytic infiltrates and lack of mitotic activity. Immunohistochemically the tumor was completely negative for PAX8 and CK7, positive for CD34 and vimentin and showed an abundant mast cell infiltrate in the CD117 staining. A staining for renin showed a strong cytoplasmic granular positivity (Figure 2).

Discussion

The juxtaglomerular cell tumor secretes excessive renin, thereby leading to secondary hyperaldosteronism and the clinical presentation with hypertension and hypokalemia [6,10,11]. As mentioned above, our patient presented without hypokalemia hinting at a secondary hypertension caused by a juxtaglomerular cell tumor. Therefore, our case report underlines the

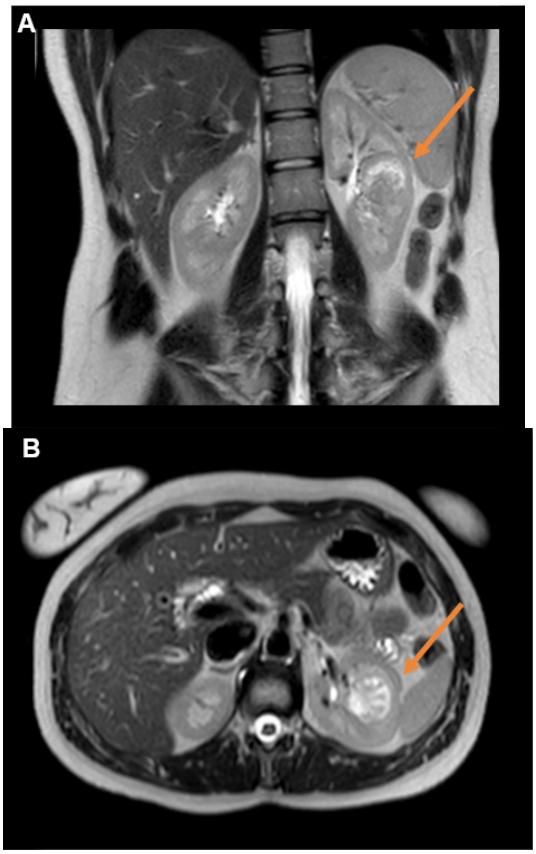

Figure 1: MRI of the abdomen showing a 32mmx29mmx27mm centrally located, solid kidney tumor in the middle pole of the left kidney (orange arrow); **A** frontal plane; **B** transversal plane.

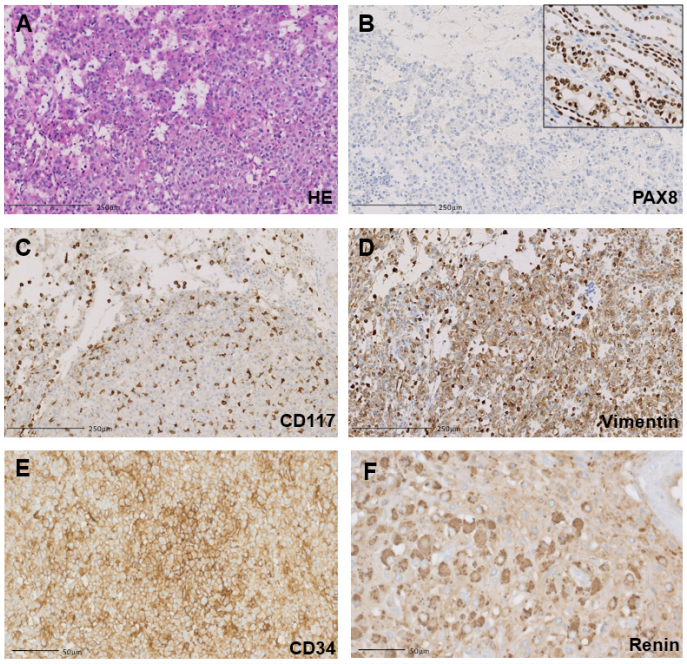

Figure 2: Histopathology of the juxtaglomerular tumor: **(A)** Glomoid appearance of slightly eosinophilic monomorphic tumor cells, **(B)** negativity for PAX8 (right upper corner renal tubules as positive control), **(C)** negativity for CD117, but abundant CD117 positive infiltrating mast cells, **(D)** slight positivity for vimentin, **(E)** positivity for CD34, **(F)** strong cytoplasmic granular positivity for renin. **(A-D)** Scale bares 250 µm. **(E-F)** Scale bares 50 µm.

diverse clinical manifestations caused by a juxtaglomerular cell tumor. Limitations of this case report include lack of ultrastructural analysis to examine the presence of renin crystals, as it has been performed by previous reports [7], but the strong renin positivity and clinical features proofed the diagnosis. Few studies have reported molecular genetics associated with juxtaglomerular cell tumors, such as monosomy of chromosomes 9 and X as well as trisomy of chromosome 10 [10]. Earlier publications mainly describe total nephrectomy as the curative therapy of choice, whereas more recent publications describe minimally invasive surgery as another therapeutic option [3,5,9].

Conclusion

Reninoma is a curatively treatable cause of malignant hypertension and kidney-sparing procedures should be the primary goal in those primarily young patients. Since juxtaglomerular cell tumors are considered benign tumors and very rarely metastasize, no mandatory follow-up is required after complete surgical resection [1, 2,12].

Statement of Ethics

There are no ethical or professional objections to the research project on the part of the Ethics Committee at the Faculty of Medicine of RWTH Aachen University. All procedures performed in this case report were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest Statement: The authors have no conflicts of interest to declare regarding this case report.

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Data Availability Statement: All data generated or analyzed during this case report is included in this article. Further inquiries can be directed at the corresponding author.

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