

Renin-secreting juxtaglomerular cell tumor of the kidney causing severe hypertension and polyuria

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Reninoma is a tumor of the juxtaglomerular cell apparatus that produces excessive amounts of renin, resulting in secondary hyperaldosteronism associated with hypertension and hypokalemia. Over the past 50 years, from 89 to 119 cases of reninoma have been reported, although in some of those cases, little clinical data have been provided.^{1,2} To the best of our knowledge, we report the first case of reninoma in Poland. We describe the clinical presentation, diagnostic workup, and management of this rare disorder.

A 31-year-old woman with a history of hypertension for 16 years, 6 months after her second childbirth, was referred to the Department of Hypertension at the Institute of Cardiology in Warsaw owing to worsening blood pressure (BP) control since delivery. The patient reported markedly increased polydipsia and polyuria (8–11 l/d), which occurred postpartum. Both her pregnancies were uncomplicated with good BP control.

On admission (while receiving methyldopa and nitrendipine), BP was 190/114 mmHg. Ambulatory BP measurement showed a mean BP of 159/106 mmHg during the day and 139/90 mmHg during the night. A physical examination showed no abnormalities. Fundoscopy demonstrated hypertensive retinopathy, grade II. Serum potassium concentration was low (2.8 mmol/l). Plasma sodium and creatinine concentrations, glomerular filtration rate, hemoglobin concentration, and platelet count were normal. Urine analysis showed no abnormalities apart from low specific urine gravity (1.002 g/ml). Echocardiography showed left ventricular concentric hypertrophy.

We performed an appropriate evaluation for identifiable causes of hypertension. Methyldopa was discontinued and the patient was treated

with doxazosin and nitrendipine during biochemical diagnostic workup. Plasma renin was 305 pg/ml (normal range, 5.1–38.7 pg/ml) and serum aldosterone was 1494 pg/ml (normal range, 35–300 pg/ml). Plasma free metanephrines, cortisol, and corticotropin were within normal ranges. Owing to persistent hypokalemia, the patient required administration of 96 mmol of potassium daily. Doppler duplex examination and computed tomographic angiography did not show any abnormalities in the renal arteries. Upper abdominal ultrasound revealed a 30-mm mass located in the right kidney. Abdominal computed tomography showed normal adrenal glands and confirmed a 30-mm mass in the upper pole of the right kidney (FIGURE 1A). Renal vein catheterization failed to show the gradient of renin concentration between the right and left renal veins.

Considering a history of recent childbirth, we performed tests to identify potential causes of polyuria. Cranial magnetic resonance imaging excluded pituitary causes of diabetes insipidus (FIGURE 1B). Hypotonic polyuria with urine osmolality of 254 mOsm/kg H₂O (normal range, 700–1200 mOsm/kg H₂O) without an increase in urine osmolality after a 3-day desmopressin test (240 µg/d) confirmed the diagnosis of nephrogenic diabetes insipidus due to hypokalemia.

We made a tentative diagnosis of a renin-producing right renal mass. The patient was put on spironolactone (50 mg/d) and enalapril (20 mg/d), which normalized serum potassium levels and improved BP control. Four weeks later, the patient underwent open partial nephrectomy of the right kidney without ischemia (FIGURE 1C). No complications were observed. Histopathology confirmed the presence of reninoma (FIGURE 1D).

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Received: February 12, 2014.
Revision accepted:
February 13, 2014.
Published online: February 20, 2014.
Conflict of interest: none declared.
Pol Arch Med Wewn. 2014;
124 (4): 207-208
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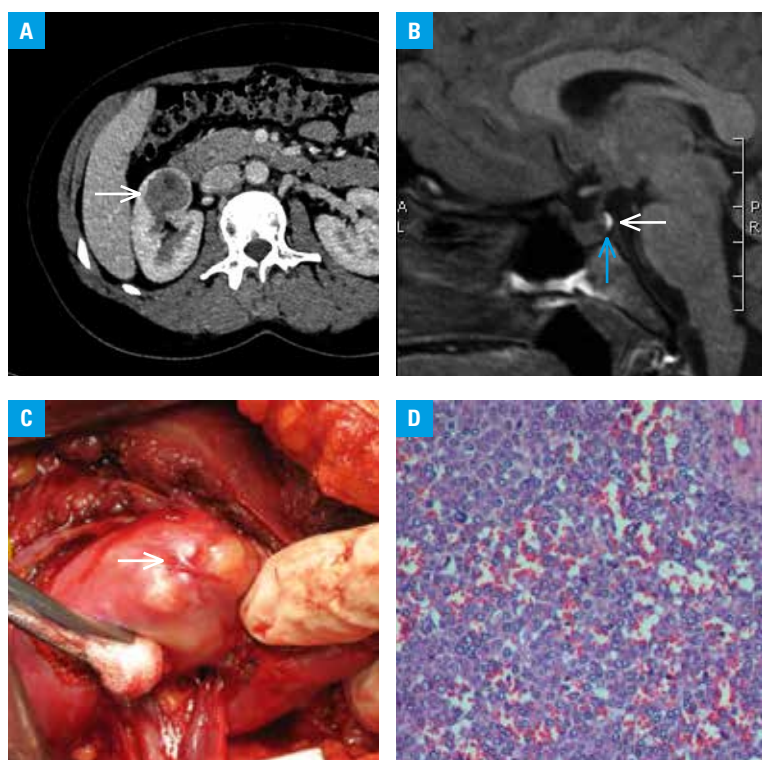


FIGURE 1 **A** – contrast-enhanced computed tomography of the juxtaglomerular cell tumor of the right kidney ($28 \times 28 \times 33$ mm); **B** – sagittal precontrast T_1 -weighted magnetic resonance image showing the pituitary gland with posterior pituitary bright spot (white arrow) and a small Rathke's cleft cyst in the intermediate lobe (blue arrow); **C** – open partial nephrectomy of the right kidney: cortical lesion (arrow) close to the renal hilum before surgical removal; **D** – pathological examination of the juxtaglomerular cell tumor

Following surgery, renin, aldosterone, and serum potassium levels normalized. Polyuria and polydipsia resolved. A month later, BP was well controlled on nitrendipine, 10 mg daily.

In summary, we reported a case of a 31-year-old patient with severe hypertension due to a reninoma in her right kidney successfully treated with partial nephrectomy. Although very rare, a renin-producing tumor should be considered in any patient with elevated renin and aldosterone levels associated with refractory hypertension, especially in the presence of hypokalemia.

Acknowledgments We would like to thank Professor Włodzimierz Januszewicz for his expertise and important suggestions during diagnostic and therapeutic steps. We would like to thank also Professor Wojciech Zgliczyński, Doctor Iłona Michałowska, and Doctor Jacek Kądziera for their expertise and help in consulting this case.

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