A 22-year-old man presented at hospital with a prolonged history of headache, polyuria and excessive thirst. His medical history included neglected hypertension since teenage. His physical exam was remarkable for a blood pressure of 190/120 mmHg. He denied any use of tobacco, alcohol, coffee, street drug and liquorice. Biochemical evaluation revealed hypokalemia at 2.9 mmol/L, hyponatremia at 134 mmol/L and normal estimated glomerular filtration rate. Twelve-lead ECG demonstrated left ventricular hypertrophy and ambulatory blood pressure monitoring confirmed severe hypertension with a dipper profile (150/100 mmHg with 50 mg of nicardipine twice daily). Transthoracic echocardiography showed moderate left ventricular hypertrophy, a LVEF of 45% and no evidence of aortic coarctation. Carotid-femoral pulse wave velocity was 8.2 m/s (reference value 8.4 m/s) and central pressure evaluated with SphygmoCor device was 134/90 mmHg (Augmentation index 17%). Twenty-four hours
Urinary analysis was consistent with a low proteinuria (0.54 g/24 hours), a normal catecholamine and cortisol levels. Doppler study with a trained physician ruled out any renal artery stenosis. Laboratory tests performed after correction of hypokalemia demonstrated a secondary aldosteronism with a marked elevation of renin and prorenin. In supine position, levels were respectively 161 pg/mL for renin (normal < 3.5 pg/mL), 2173 pg/mL for prorenin (normal < 135 pg/mL), 557 pg/mL for aldosterone (normal < 105 pg/mL) and aldosterone-to-renin ratio was 3.5 (normal < 46.9). After one hour of standing position renin and prorenin levels were similar (166 pg/mL, normal value < 7.2; 2123 pg/mL, normal range < 150, respectively) conversely with a three-time increase of aldosterone (1565 pg/mL, normal < 275) and aldosterone-to-renin ratio at 9.4 (normal < 46.9).

Twenty four hours aldosteronuria was 48.4 μg (normal < 32). Abdominal computed tomography showed the presence of a 3 cm mass in the upper pole of the left kidney without enhancement (figure 1). Magnetic resonance imaging with and without gadolinium confirmed a well-defined hypointense mass lesion with no contrast enhancement (figure 2). Selective renal sampling was performed to determine the function of this lesion. There was a two-time gradient of renin and prorenin in the left renal vein (271 pg/mL, 3219 pg/mL respectively) compared to the right one (140 pg/mL, 1731 pg/mL respectively).

What is your diagnosis?
All these data taken together, the diagnosis of reninoma was considered. The patient underwent laparoscopic tumorectomy to remove the cortical lesion. Pathology examination confirmed a well-encapsulated lesion measuring 3 cm without sign of malignancy. Immuno-histochemistry study demonstrated a typical pattern of reninoma: antibodies were positive for renin, vimentin and CD34. His postoperative course was uneventful and he was discharged 7 days after surgery. One month after discharge, the patient was asymptomatic and ambulatory blood pressure monitoring without antihypertensive treatment showed values at 115/75 mmHg.

When the onset of hypertension started during childhood, more than a half of cases are related to secondary hypertension. Main etiologies are renal parenchymal disease, coarctation of the aorta, fibromuscular dysplasia, thyroid dysfunction and few decades later Cushing syndrome, primary aldosteronism, obstructive sleep apnea, pheochromocytoma and Takayasu’s disease [1,2]. Secondary aldosteronism should be suspected in the presence of hypokalemia and slightly low natremia, conversely natremia is usually slightly high in primary aldosteronism. When the hormonal profile confirmed a secondary aldosteronism, diagnosis approach must consider either a renal perfusion defect (coarctation of aorta, renal artery stenosis, fibromuscular dysplasia, ischemic kidney….) or an inappropriate secretion of renin from an endocrine tumor (reninoma, teratoma, paraneoplastic syndrome….) [3,4].

Reninoma is an extremely rare tumor of the renal juxtaglomerular cell apparatus, which induced a high plasma level of renin. A recent literature review reported 89 cases of reninoma [4]. The mean age at diagnosis was 27 years. A half of patients suffered from headache and mean blood pressure was 201/130 mmHg. Hypokalemia was detected in more than 80% of cases. Plasma renin activity and aldosterone levels were respectively 12 and 4 four times the upper limit of normal. In our report, the renin/total renin ratio was 7.4% in the peripheral vein, 8.4% in the left renal vein and 12.4% in the right renal vein. These ratios were below the value of normotensive young white subjects (range from 17.1 to 22.9), which suggests a down regulation of prorenin proteolysis in reninoma [5]. CT and MRI are both good tools to detect the tumor, which sized a mean diameter of 3 cm. Renal vein catheterizations emphasized in a half of cases the presence of lateralization of renin secretion. According to location of the tumor, radical or partial nephrectomy was performed. Pathology usually confirmed diagnosis in presence of immuno-histochemical stains for renin, actin, vimentin and CD34. Around 90% of subjects were cured of hypertension after surgery.

To conclude, reninoma is an uncommon cause of secondary aldosteronism. In the absence of other diagnosis related to this hormonal status (coarctation of aorta, renal artery stenosis, fibromuscular dysplasia, ischemic kidney….), reninoma must be ruled out because hypertension can be definitely cured with a specific management.

Disclosure of interest: the authors declare that they have no conflicts of interest concerning this article.

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