Gordon's Syndrome – Hypertension and Hyperkalemia Associated with Normal Glomerular Filtration Rate

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Dear Sir,

A constellation of hyperkalemia and arterial hypertension associated with intact renal function was first described by Gordon et al. [1]. Until 1986, 28 patients with this syndrome were documented. Less than half of them were single cases, while the remainder were members of two families from Israel and the USA [2].

A 36-year-old man with arterial hypertension (with a maximum value of 180/130 mm Hg) and hyperkalemia (in the range of 5.5–6.5 mmol/l) was admitted in the clinic. The patient was free of noteworthy complaints and coped successfully with his duties as a driver. The physical examination revealed no pathological changes. Blood pressure was between 170/110 and 140/90 mm Hg untreated. There was no deviation in the essential clinical chemistry data. The repeated assessment of serum electrolytes disclosed values as follows: potassium, 5.6–7.1 mmol/l; sodium, 140–146 mmol/l; chlorides, 104–118 mmol/l; calcium, 2.8 mmol/l, and magnesium, 0.7 mmol/l. The clearances of the basic electrolytes yielded the following results: potassium, 0.06 ml/s (at norm 0.5); sodium, 0.017 ml/s (at norm 0.17); chlorides, 0.01 ml/s (at norm 0.0117–0.067); acid-base balance was pH 7.31, BE/ 4.9 mmol/l; creatinine clearance, 1.88 ml/s; plasma renin activity in the morning in recumbency, 0.17 pmol/l (at norm 0.043–0.578); aldosterone, 0.4 nmol/l (at norm 0.136–0.540); cortisol, 448 nmol/l (at norm 165–632). EKG showed hyperkalemic T waves. Computed tomography of adrenals and X-ray study of sella turcica demonstrated no pathological changes. Kidney biopsy showed that the investigated 16 glomeruli were totally intact. The arterioles presented advanced changes of hypertonic type.

The patient was given treatment with nife-dipin and furosemid at a daily doses of 40 mg each, resulting in blood pressure from 140/90 to 150/95 mm Hg and serum potassium from 5.9 to 4.7 mmol/l.

The observation presented has aroused considerable interest because it combines all clinical signs characterizing the extremely rare syndrome of Gordon, hyperkalemia with normal glomerular filtration rate, arterial hypertension, very good response to diuretic treatment (in our case combined with nifedipine), leading to serum potassium level and blood pressure normalisation. In the case reported on, there were no data pointing to hemolysis, infectious

mononucleosis, leuko-cytosis and thrombocytosis that would eventually condition pseudohyperkalemia.

The pathogenetic mechanisms of the syndrome are still not well clarified. No deviations whatsoever were recorded in plasma renin activity and serum aldosterone level, and therefore, we could hardly lend support to the hypothesis that sodium retention is the underlying cause of the decrease in renin level with ensuing deficient aldosterone production [1].

The presumption of a genetic defect in the membrane of tubular cells with disorders in potassium transport [3] seems more likely.

References

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