Pseudohypoaldosteronism type 2 (PHA2), also known as Gordon syndrome, is a rare inherited form of low-renin hypertension associated with hyperkalaemia and hyperchloremic metabolic acidosis in patients with a normal glomerular filtration rate (GFR). PHA2 is the result of mutations in a family of serine-threonine kinases called with-no-lysine kinases (WNK) 1 and WNK4. These enzymes regulate electrolyte channels in the aldosterone sensitive distal nephron, resulting in decreased potassium secretion. It is hypothesized that patients with PHA2 have a chloride shunt – increased distal chloride reabsorption, leading to less electronegativity in the lumen, thereby decreasing the electrochemical gradient for potassium secretion.

Some causes of low renin hypertension

<table>
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<tr>
<th>Primary hyperaldosteronism</th>
<th>Pseudohypoaldosteronism</th>
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<td>Macrovascular adrenal hyperplasia</td>
<td>Apparent mineralocorticoid excess (HSD-2 deficiency)</td>
<td>Pseudohypoaldosteronism type 2 s. Gordon syndrome</td>
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<td>Aldosterone-producing carcinoma</td>
<td>Mineralocorticoid receptor mutation</td>
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A 57 year old female patient with known adrenal mass, a typical adenoma found on abdominal CT 5 years ago with the diameter of 1.7 cm

Laboratory test results excluded Cushing syndrome, but primary hyperaldosteronism was not suspected due to persisting hyperkalaemia

She had anamnesis of treatment-naïve hypertension, cardioembolic stroke in 2011 and foramen ovale apertum enclosed with occluder in 2012

Abdominal CT scan was repeated to evaluate the growth dynamics of the adrenal mass, revealing an enlarged left adrenal gland with a hypodense (-9 HU) vascular mass 4.1 cm in diameter

Admitted to the hospital for left adrenalectomy

Pre-operative testing revealed several laboratory test abnormalities

Considering the findings of low renin hypertension with hyperkalaemia and normal GFR the diagnosis of PHA2 was established.

The patient was started on antihypertensive treatment with thiazide diuretics and underwent left adrenalectomy.

Histology confirmed a hormonally inactive adenocortical adenoma, Weiss score 2.

CONCLUSIONS

The current case demonstrates the challenges of differential diagnosis of low renin hypertension depicting the characteristic findings of PHA2. It should be noted that upon correct diagnosis this form of low renin hypertension is easily treatable with thiazide diuretics.

References


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