Challenges in the diagnosis of pheochromocytoma - a case report

Aleksandra Zdrojowy-Wełna, Grażyna Bednarek-Tupikowska
Department of Endocrinology, Diabetology and Isotope Therapy, Wrocław Medical University

Introduction

Adrenal pheochromocytomas are rare neuroendocrine tumours, however their prevalence is probably underestimated – in some series 50% were diagnosed at autopsy. The clinical presentation varies among patients, that is why diagnosis might be delayed.

Case report

49-year old woman with hypertension was referred to the Endocrinology Department in Wrocław with suspected pheochromocytoma.

2002: operation of right carotid paraganglioma

2012: operation of left carotid paragangioma, complicated with stroke

- administration of anticoagulants
- bleeding from gastrointestinal tract
- CT of abdomen:
  - tumour in the right adrenal gland 6,5 x 5,5 x 4,3 cm
  - tumour in left ovary 14 x 18 x 7 cm

Clinical symptoms: uncontrolled hypertension with episodes of hypotension, weight loss (11 kg in 2 months), diabetes

Family history:
- sister - carotid paragangioma
- father - cardiovascular disease (†56yr)

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<tbody>
<tr>
<td><strong>Adrenaline in blood</strong></td>
<td>10,64 nmol/l [&lt;0,69]</td>
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<tr>
<td><strong>Noradrenaline in blood</strong></td>
<td>20,5 nmol/l [&lt;3,55]</td>
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<tr>
<td><strong>Adrenaline in urine</strong></td>
<td>1230 nmol/l [&lt;110]</td>
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<tr>
<td><strong>Noradrenaline in urine</strong></td>
<td>3423 nmol/l [&lt;535]</td>
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<td><strong>Metoxycate cholamines in urine</strong></td>
<td>4503 ug/24h [&lt;1000]</td>
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The patient was operated on January 2013 after administration of alfa- and beta-blockers. Histopathological examination:

**pheochromocytoma epinephri**: IKi67 (+)<1%, synaptophysin (+), chromogranin A (+), S100 (+), CD56(-), Melan A (-), alfa 1 inhibin (-), CD34 (-), Calretinin (-)

**teratoma maturn cysticum ovarii**

After removing pheochromocytoma most of the clinical symptoms subsided.

Genetic screening mutation in SDHD gene positive (*)

Diagnosis: pheochromocytoma-paragangioma syndrome

Conclusions

1. Occurrence of paragangioma with hypertension suggest need of screening for pheochromocytoma-paragangioma syndrome, especially in case of paragangiomas in family history.

2. Early treatment is crucial to avoid life-threatening cardiovascular complications.