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Authors: Anna Kurzyńska, Anna Sowa-Staszczak, Andrzej Budzyński, Magdalena Ulatowska-Białas, Alicja Hubalewska-Dydejczyk, Elwira Przybylik-Mazurek

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Imaging possibilities in Cushing's ectopic syndrome due to pheochromocytoma may help to avoid delayed optimal treatment in atypical clinical course

Anna Kurzyńska¹, Anna Sowa-Staszczak¹, Andrzej Budzyński², Magdalena Ulatowska-Białas³, Alicja Hubalewska-Dydejczyk¹, Elwira Przybylik-Mazurek¹

¹ Department of Endocrinology Medical College Jagiellonian University Krakow, Poland,
² II Chair of Surgery Medical College Jagiellonian University Krakow, Poland,
³ Chair of Pathomorphology Medical College Jagiellonian University Krakow, Poland.

**Corresponding author:** dr hab n. med. Elwira Przybylik–Mazurek

Department of Endocrinology, Medical College Jagiellonian University Krakow, Poland.

ul. Mikołaja Kopernika 17

31-501 Kraków

Tel. + 48 12 424 75 20

Email: elwira.przybylik-mazurek@uj.edu.pl

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Cushing’s syndrome due to ectopic ACTH (adrenocorticotropic hormone) secretion (EAS) is a rare endocrine condition responsible for about 5–20% of all cases of Cushing’s syndrome (CS) [1-4]. In this group, pheochromocytoma is the source of ACTH only in about 5% of cases [5]. We present two clinical cases of patients with EAS caused by pheochromocytoma. The first patient was 70-year-old female with a history of hypertension, hyperlipidemia and ischemic heart disease and type 2 diabetes mellitus. The patient reported that she had been experiencing fatigue and muscle weakness for several months as well as decreased appetite and weight loss of 10 kg within 3-4 months. About two months earlier she has been hospitalized with suspected acute coronary syndrome. Coronarography did not reveal significant abnormalities, but it was complicated by an acute post-contrast kidney injury (AKI). Because of mental confusion, head imaging was performed: partially empty Turkish saddle was suspected based on the results of CT (computed tomography), which was confirmed in MRI (magnetic resonance imaging) of the pituitary gland. At admission, she was in superficial verbal contact, with limited auto and allogenic orientation, BP (blood pressure) 115/70 mmHg, edema of the legs, muscle atrophy, thin, dry skin with a diffuse hyperpigmentation, multiple petechiae and ecchymosis on the skin were present. Laboratory data are presented in Table 1. EAS caused by pheochromocytoma was suspected based on the clinical outcome and the diagnostics performed so far. Because of the recent AKI episode, the patient was not referred to abdominal CT scan as first-line imaging examination. Somatostatin receptors imaging (SRI) with Ga-68 labelled somatostatin analogue was performed and the accumulation of a tracer in the right adrenal gland was observed (Figure 1A). MRI of the abdomen confirmed tumor of the right adrenal gland, measuring 44x41x36mm (Figure 1B). After pharmacological preparation the patient underwent a right-side laparoscopic adrenalectomy. Histopathological examination confirmed pheochromocytoma with
chromogranin A and focal expression of ACTH-secreting cells (Figure 1C, Figure 1D).

Unfortunately, on the 9th day after surgery the patient died of ARDS.

The second patient was a 61-year-old female with a 2-month history of weight gain, proximal myopathy, depressive disorders, abdominal pain, diabetes mellitus of recent onset and worsening control of hypertension. Physical examination revealed facial and leg edema, plethoric face, dermal and muscle atrophy and moderate central obesity. Laboratory data are presented in Table S1. Abdominal CT scan and SRI showed 30 mm mass in the right adrenal gland, with radiological suspicion of pheochromocytoma (Figure 1E, Figure 1F). After pharmacological treatment, the patient was successfully operated laparoscopically. Histopathological examination confirmed pheochromocytoma with focal expression of chromogranin A and ACTH-secreting cells (Figure 1G, Figure 1H). Currently, the patient is in a complete remission and does not require any treatment.

EAS caused by pheochromocytoma is extremely rare, but should be considered as a possible source of ACTH production as it is a life-threatening condition if it is not diagnosed early and treated properly. A longer history and a cardiac or renal burden are associated with a worse prognosis.
References:


Figure 1. **A** Somatostatin receptors imaging (SRI) with Ga-68 labelled somatostatin analogue. Arrow shows accumulation of a tracer in the right adrenal gland of the Patient 1 (Department of Endocrinology Medical College Jagiellonian University Krakow, Poland). **B** Magnetic resonance imaging of the abdomen demonstrating right adrenal pheochromocytoma of Patient 1 - coronal image. Arrow shows large heterogeneous mass, measuring 44x41x36mm. (Department of Diagnostic Imaging University Hospital in Krakow, Poland). **C** Histological examinations of the Patient1. Chromogranin- A (Chair of Pathomorphology Medical College Jagiellonian University Krakow, Poland). **D** Reaction to ACTH of the Patient1 (Chair of Pathomorphology Medical College Jagiellonian University Krakow, Poland). **E** Abdominal computed tomography (CT) of the Patient 2. Arrow shows 30 mm mass in the right adrenal gland with radiological suspicion of pheochromocytoma. (Department of Diagnostic Imaging University Hospital in Krakow, Poland). **F** Somatostatin receptors imaging (SRI) with 99mTc labelled somatostatin analogue Arrow shows accumulation of a tracer in the right adrenal gland of the Patient 2 (Department of Endocrinology Medical College Jagiellonian University Krakow, Poland). **G** Histological examinations of the Patient 2. Chromogranin- A (Chair of Pathomorphology Medical College Jagiellonian University Krakow, Poland). **H** Reaction to ACTH of the Patient 2. (Chair of Pathomorphology Medical College Jagiellonian University Krakow, Poland).