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Difficulties in the diagnosis and treatment of ruptured pheochromocytoma

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We present a case of 68-year-old patient, who was admitted to the Emergency Department with severe abdominal pain and nausea. Moreover, the patient had been vomiting repeatedly for the past 2 days, and he had suffered from tachycardia, dyspnoea on exertion, and deterioration of general condition over the past 2 weeks. The patient had a past medical history of hypertension, but apart from that he had no other medical conditions. On admission, paroxysmal hypertension up to 240 mm Hg of systolic blood pressure, followed by hypotension (about 60/40 mm Hg) and an episode of seizures with concomitant apnoea lasting 10 minutes, were observed. The patient needed active ventilation with an Ambu device providing 100% oxygen, which resulted in a return to spontaneous breathing. Computed tomography angiography (angio-CT) excluded pulmonary embolism; however, it showed a right adrenal mass (size 110 × 65 × 80 mm) with heterogeneous enhancement and suspicion of rupture of the central part of the tumour with contrast extravasation to intraperitoneal and retroperitoneal space (Fig. 1). In the left adrenal gland, another mass, 14 mm in its largest dimension, was observed. The patient was urgently referred to the Surgical Department. Upon admission, he was haemodynamically stable with preserved spontaneous breathing. Additionally, higher blood pressure up to 160/90 mm Hg was noted. His level of cortisol was adequate to his condition (23.5 ug/dL). The serial complete blood count monitoring (CBC) showed a rapid decrease of the haemoglobin level (from 9.3 g/dL to 7.9 g/dL) shortly after admission. Due to of suspicion of active bleeding from a ruptured tumour

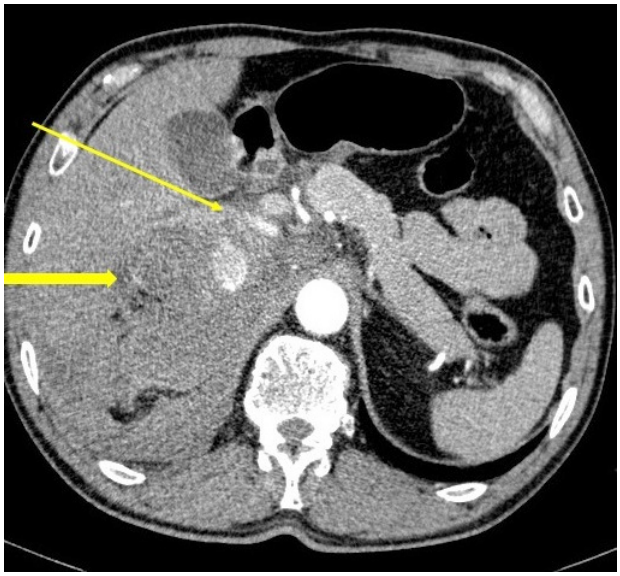

Figure 1. *Computed tomography angiography (angio-CT), arterial phase, axial image — right adrenal mass with heterogeneous enhancement (thick arrow), suspicion of rupture of central part of the tumour with contrast extravasation to retroperitoneal space (thin arrow). Imaging result obtained from the Department of Diagnostic Imaging, Hospital in Sucha Beskidzka, Poland*

Table 1. ***The results of 24-hour urine collection of methoxycatecholamines after the operation***

Normetanephrine	176.9 ug/24 h (88–440)
Metanephrine	27.8 ug/24 h (52–341)
3-Metoksytyramine	76.7 ug/24 h (0–220)

Urine volume: 2000 mL

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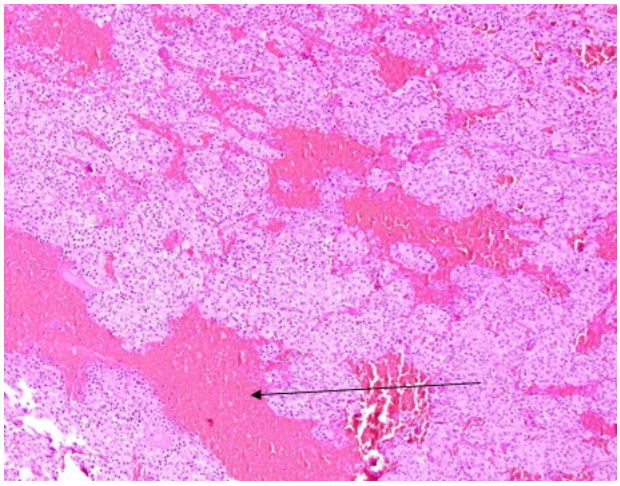

Figure 2. *Central part of the pheochromocytoma with irregular, diffuse haemorrhages between tumour nests (arrow)*

and a high risk of haemodynamic instability, the patient was qualified for live-saving surgery. Therefore, based on the clinical symptoms and radiological features, suspicion of the ruptured haemorrhagic pheochromocytoma existed and pharmacological treatment with alpha-receptor blockers and fluid infusions was started without performing 24-hour urine fractionated metanephrine collection. The patient received gradually increasing doses of doxazosin for the next 4 days at up to 6 mg per day. He also required transfusion of 3 units of packed red blood cells. Afterwards, an urgent right adrenalectomy was performed. No peri-operative and post-operative complications were observed. Histopathological examination confirmed haemorrhagic pheochromocytoma in the right adrenal gland (Fig. 2). There were massive haemorrhagic changes in the lesion, proliferation index Ki-67 was 3.4%, and the Pheochromocytoma of the Adrenal gland Scaled Score (PASS) score was 2, suggesting a benign character of the neoplasm. A small abscess was found near the tumour capsule. After the operation, the patient remained stable and in good clinical condition. Doxazosin was gradually titrated and finally withdrawn a few days later. Postoperatively, both 24-hour urine fractionated metanephrines, cortisol, and chromogranin A concentrations were within the normal range. A CT scan 12 months after the first one showed no progression of the left adrenal adenoma. Currently, the patient remains under the care of the Endocrinology Department at the University Hospital in Cracow with a follow-up period of 2 years, without any evidence of disease recurrence. He remains in good clinical condition, without alarming symptoms. The patient uses only a low dose of angiotensin receptor blocker for mild hypertension. A 24-hour urine collection of methoxycatecholamines showed normetanephrine, metanephrine, and 3-methoxythyramine concentrations within the normal range — the results excluded pheochromocytoma in the left adrenal gland. Genetic testing for *RET*, *VHL*, *SDHB/SDHD*, *MAX*, and *MEN1* mutation was negative.

We present a case of adrenal bleeding, which was the first manifestation of pheochromocytoma. Spontaneous adrenal haemorrhage associated with a mass is uncommon, and treatment strategies have not been standardized. When it occurs, malignant disease or pheochromocytoma should be excluded in the first place. In the literature, pheochromocytoma is presented as the most common cause of adrenal bleeding with an accompanying mass, and it accounts for nearly 50% of cases. When a ruptured pheochromocytoma is suspected, abdominal CT scan is a gold standard. Typically, heterogeneously enhanced mass and haematoma in the suprarenal region of the retroperitoneum are obtained. Usually patients present acute abdominal, lumbar, or chest pain. We can also observe accompanying hypertensive attacks, headaches, palpitation, and hidrosis. Poorly controlled preoperative blood pressure is a risk factor for perioperative morbidity, but intraperitoneal bleeding is an indication for emergency exploratory laparotomy. Adrenalectomy should be performed on stable patients after hormonal assessment and preoperative treatment by alfa-blockers. Embolization for ruptured pheochromocytomas may be preferred as the initial therapy because emergency surgery carries a mortality rate of 45%. It can be helpful in stabilizing the patient's condition and it can give some time to properly prepare the patient for the elective surgery. Awareness of the possible manifestation of a haemorrhagic pheochromocytoma is very important for the management of this rare but lethal condition.

Author contributions

The first authorship of J.K and E.R is of equal rank.

Conflict of interest

None declared.

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