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CASE REPORT

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Multidrug-resistant hypertension. A case report.

Wielolekooporne nadciśnienie tętnicze. Opis przypadku.

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Abstract

Pheochromocytoma is a rare, mostly benign catecholamine-producing tumor that arises from chromaffin cells of the adrenal medulla. In most cases the symptoms are not specific for this disease. Multidrug-resistant hypertension could be one of them.

In this case report, we present a patient who had been treated for years because of hypertension without any response to treatment. During the diagnostic process pheochromocytoma have been found and the proper treatment had been provided resulting in good outcomes.

It should be noted that not only the clinical symptoms, but also the planning of an appropriate diagnostic in close cooperation with the radiologists, is very important during the whole diagnostic.

Key words: *Pheochromocytoma, multi-drug resistant hypertension, radiology, CT, MRI*

Streszczenie:

Guz chromochłonny jest rzadkim, najczęściej łagodnym guzem produkującym katecholaminy, który powstaje z komórek chromochłonnych rdzenia nadnerczy. W większości przypadków objawy guza nie są specyficzne dla tej choroby. Nadciśnienie tętnicze wielolekooporne może być jednym z objawów pheochromocytoma.

W tej pracy przedstawiono przypadek pacjenta, który od lat był leczony z powodu nadciśnienia tętniczego bez żadnej poprawy. Podczas prowadzonej diagnostyki wykryto guza chromochłonnego i wdrożono odpowiednie leczenie z dobrym efektem.

Należy pamiętać, że nie tylko objawy kliniczne, ale również zaplanowanie odpowiedniej diagnostyki w ścisłej współpracy z radiologiem, jest bardzo ważne podczas całej prowadzonej diagnostyki.

Słowa kluczowe: *Pheochromocytoma, wielolekooporne nadciśnienie tętnicze, radiologia, tomografia komputerowa, rezonans magnetyczny*

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Introduction

In the current case report, we present a case of a patient who has been diagnosed with a drug-resistant hypertension.

Hypertension is the most common chronic disease and the leading risk factor for disability.

Multidrug-resistant hypertension is a particularly severe form of hypertension, one of the causes of which is pheochromocytoma.

A pheochromocytoma is a rare, mostly benign catecholamine-producing tumor that arises from chromaffin cells of the adrenal medulla. It is estimated that the annual incidence of pheochromocytoma is approximately 0.8 per 100.000 person-years. In most cases this tumor occurs sporadically, however it can be associated with genetic syndromes such as Von Hippel-Landau (VHL) disease, type 1 Neurofibromatosis and Multiple Endocrine Neoplasia type 2 (MEN2) in 10-25% of the cases [1]. Pheochromocytoma is characterised by a highly variable clinical presentation, but most commonly presents with episodes of headaches, sweating, palpitations and hypertension [2].

Hypertension is often paroxysmal in nature. Diagnostic imaging is required to settle the diagnosis and plan the therapeutic process. Laparoscopic tumor removal is now the gold standard for the tumor therapy [3]. If removal is timely, prognosis is excellent (in benign pheochromocytomas, the 5-year survival rate is above 95%) [3,4].

Case presentation

A 63-year-old patient was admitted to the Clinical Department of Gastroenterological Surgery and Transplantation of the Central Clinical Hospital of the Ministry of Interior and Administration in Warsaw in June 11, 2021 due to epigastric pain and a 4-day long episode of diarrhea 2 weeks prior to admission. The symptoms were not related to any obvious cause.

The patient denied sweating, headaches, palpitations, tremors, constipation, nausea and vomiting. Physical examination showed a heart rate of 84 beats/min and blood pressure of 150/90 mmHg. His history included long lasting hypertension treated with a calcium channel blocker (Lacipil) and angiotensin-II receptor blocker (Telmisartan). Since the diagnosis was settled intermittent increases in blood pressure occurred (recorded SBP 210mmHg 20 years ago and SBP 180 mmHg during quarantine due to suspected COVID). The patient was taking antihypertensive medications irregularlydue to the tendency to low blood pressure in home measurements. The patient's family history did not include neoplastic diseases, diseases of the thyroid gland nor adrenal glands.

The diagnostic pathway of the multidrug-resistant hypertension included blood tests and the abdominal ultrasound examination followed by CECT of the abdomen. Abdominal CECT scan was performed on June 11, 2021. It revealed a heterogenous, strongly enhancing focal lesion of the right adrenal gland (Figure 1). The pattern of contrast enhancement and the morphology of the lesion indicated a pheochromocytoma. Computed tomography examination enabled settlement of the cause of multidrug-resistance hypertension and localization of the lesion, which is crucial for the surgical treatment of the tumor.

Biochemical tests showed increase concentration and daily excretion of metanephrines (concentrations in consecutive measurements 358/247/220 µg/l with normal up to 145 µg/l; excretion 1074/693/772 µg/24h with normal up to 224 µg/24h; daily urine collection 3000/2800/3500 ml). There was no evidence for MEN syndromes and hyperparathyroidism in biochemical tests. Considering the patient's clinical signs including hypertension with intermittent increases in blood pressure, as well as his laboratory tests and clinical imaging report, the diagnosis of right adrenal pheochromocytoma was settled. The patient was qualified for right-sided adrenalectomy. Treatment with oral selective antagonist of alfa1-adrenergic receptors (Doxasozin in a gradually increasing dose, till the end to 16 mg per day before the surgery) and Ca channel blocker (10 mg per day) was initiated to normalize BP. After exclusion of surgical contraindications, the patient underwent surgery on July 14, 2021. During the procedure the tumor of the right adrenal gland was removed (Figure 2). Histopathological evaluations confirmed the diagnosis of pheochromocytoma. After the surgery, blood pressure was constantly monitored. The patient was discharged 7 days after the surgery with normalised blood pressure.

Discussion

Adrenal pheochromocytoma is known as a rare tumor arising from catecholamine-producing chromatin cells. The most accurate diagnostic test for the biochemical diagnosis is the measurement of plasma free or 24-hour urinary fractionated metanephrines. Although the typical symptoms and signs of pheochromocytoma such as headache, tremors, palpitations, sweating and hypertension are well known, up to 25% of patients do not present signs and symptoms, and up to 30% pheochromocytomas are diagnosed following the discovery of an adrenal incidentalo-

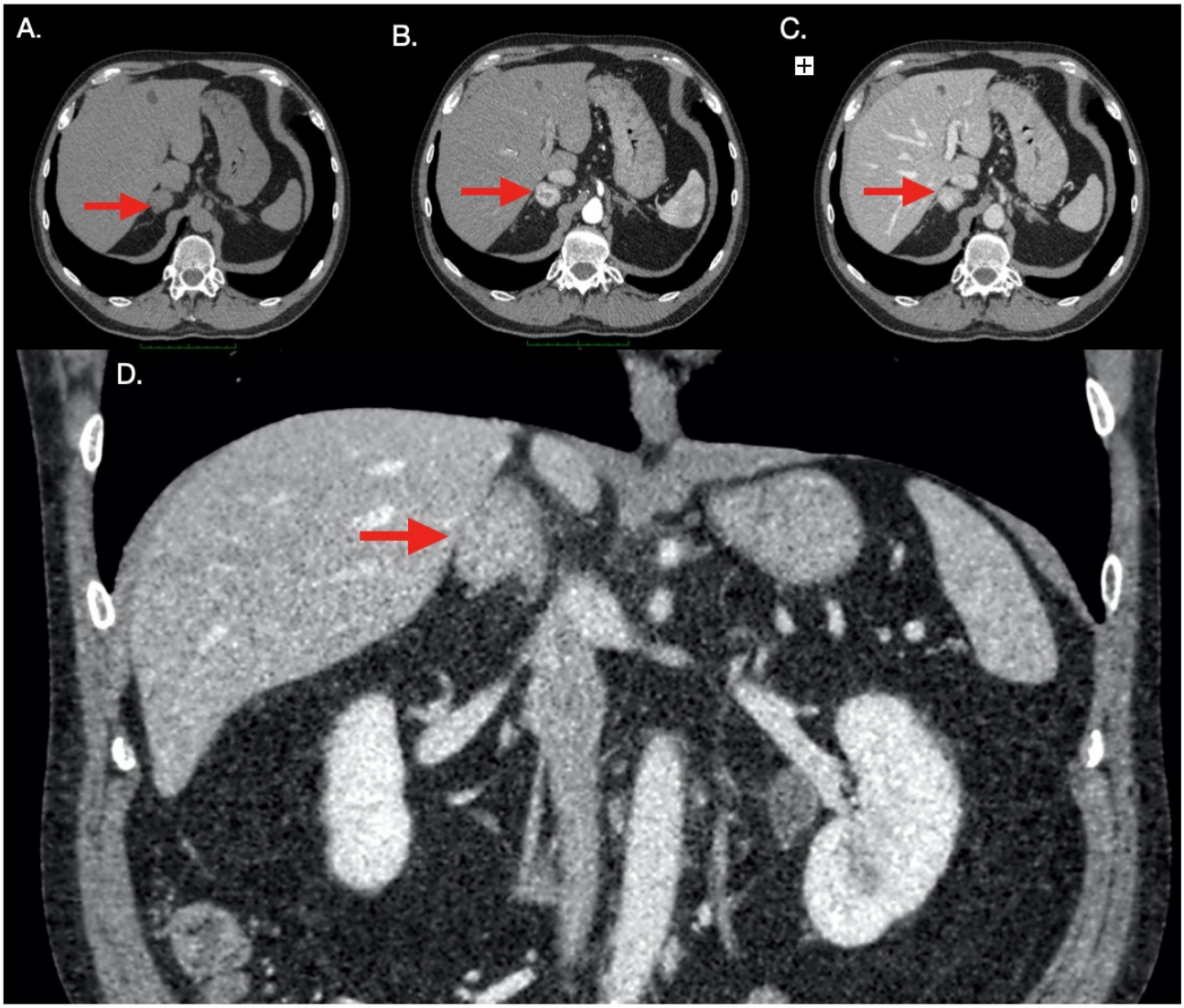

Figure 1 *Abdominal CT scan. The lesion of right adrenal gland is pointed with an arrow: A - native (non-contrast) phase - lesion isodense; B - arterial phase - strong contrast enhancement; C, D - venous phase - sustained contrast enhancement. The tumor density was above +10 HU and it had a low contrast washout rate. These features are suggestive of pheochromocytoma.*

ma [5]. According to European Society of Endocrinology Clinical Practice Guideline in collaboration with the European Network for the Study of Adrenal Tumors, pheochromocytomas account for up to 7% of adrenal incidentalomas [6].

Missed diagnosis and misdiagnosis of the cause of multidrug-resistant hypertension can happen easily. It is extremely important to think about pheophromocytoma during the diagnostic process of hypertension. Pheochromocytoma as a cause of arterial hypertension should be differentiated from renal artery stenosis (RAS). The most common causes of stenosis among adults are atherosclerotic lesions, which are responsible of 85-90% of cases [7]. The most important diagnostic methods in RAS are Doppler ultrasonography (USG), computed tomography (CT), magnetic resonance imaging (MRI) [8]. If RAS is excluded retroperitoneal space should be examined. The diagnostic value of ultrasound is insufficient since the visibility of the retroperitoneum is poor and the differentiation between incidentalomas and pheochromocytomas is deteriorated. The diagnosis of pheochromocytoma and its localization is usually based on CT and/or MRI imaging. The sensitivity of CT scans reaches 85-94% and the specificity 92% [4]. The greatest advantage of computed tomography versus MRI are cost-effectiveness and high sensitivity. On the other hand, MRI provides superior contrast resolution in soft tissues and therefore better differentiation of pheochromocytoma from adrenal adenomas and also allows exclusion of vascular invasion. Moreover, MRI does not use radiation [4].

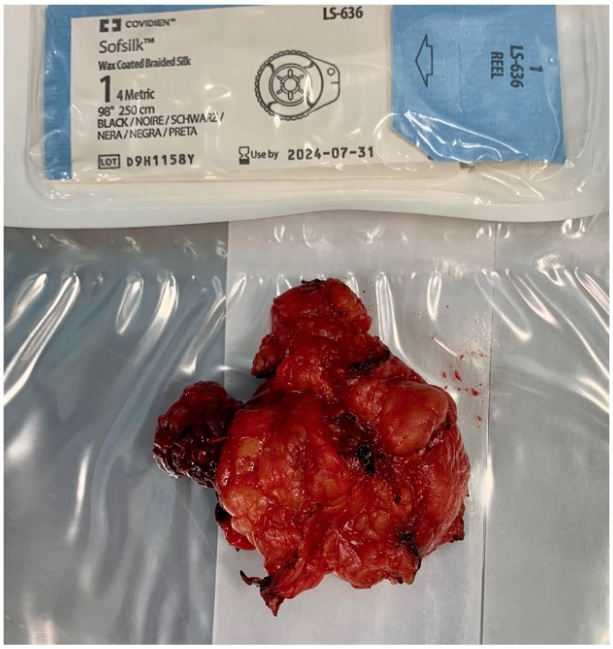

Figure 2 *The tumor of the right adrenal gland removed during the surgery*

All in all, early and accurate diagnosis of pheochromocytoma is crucial to the patient. Therefore, it is worth remembering that performing of the CT or MRI is necessary to state the proper diagnosis and the close cooperation with radiologist is important during the whole diagnostic and treatment.

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