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PHEOCROMOCYTOMA IS A RARE TUMOR WITH A DIFFICULT DIAGNOSIS THAT USUALLY CAUSES HYPERTENSION

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ABSTRACT

Pheochromocytoma is a rare neuroendocrine tumor, with a prevalence rate of less than 0.1%, that is difficult to diagnose and treat. The tumor originates from the chromaffin cells of the adrenal gland and affects women more than men. The diagnosis is often confused with other disorders such as preeclampsia, eclampsia, and thrombocytopenia. The disease causes secondary hypertension that can be followed by stroke, myocardial infarction, and death, and mechanical effects such as palpation of the abdomen at the adrenal glands can cause paroxysmal seizures. In patients suffering from this disease, the skin may appear light brown and this sign can help with diagnosis. Headache, sweating, and especially hypertension are important diagnostic indicators for pheochromocytoma. Hyperglycemia, hypercalcemia, proteinuria, and the presence of lactic acid in the blood can also be diagnostic aids. Routine clinical tests may include catecholamine and urine measurements, and other diagnostic tests utilizing magnetic resonance imaging (MRI) and computerized tomography (CT), and scintigraphy allow the localization of the tumor.

KEYWORDS: *pheochromocytoma, diagnosis, neuroendocrine, tumor, hypertension*

INTRODUCTION

Neuroendocrine tumors are very rare and can be benign or malignant with different pathogenicity (1). Their heterogeneity causes difficulty with diagnosis and therapy (2). Pheochromocytoma is a rare tumor that arises from the chromaffin cells of the adrenal gland that secrete catecholamines (3). It is present in less than 0.1% of hypertensive patients (4). Pheochromocytoma can occur at any age but is more common after the age of 30. The tumor affects women more than men, but not in childhood, where it seems to prevail slightly in the male sex, and it is more frequent between the ages of 8 and 14 (5). In pheochromocytoma patients, severe hypertension is often present, but not all patients have paroxysmal episodes (6). During pregnancy, the disease occurs in 1 case in 50,000 and often leads to maternal-fetal mortality (7). Pheochromocytoma is difficult to diagnose and can be confused with other pathologies such as preeclampsia, eclampsia, and thrombocytopenia (8).

DISCUSSION

Pheochromocytoma can produce secondary hypertension, stroke, myocardial infarction, and death (9). The diagnosis is based on the symptomatic results represented by paroxysmal attacks with headache, sweating, palpitations, anxiety,

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and chest or abdominal pain. Additionally, patients may also experience nausea, vomiting, tremor and weakness, weight loss, and reduced breathing (10).

Pheochromocytoma patients present hypertension with a frequency of over 90% and can have an episodic form of the disease with paroxysmal or stable crises (11). The evidence of orthostatic hypotension, followed by clinostat hypertension, should be highlighted (12). Palpation of the patient's abdomen in correspondence with the adrenal glands can cause paroxysmal crisis, suggesting the hypothesis of pheochromocytoma (12). There may be light brown skin patches that suggest neurofibromatosis (13). In pheochromocytoma, tachycardia can also occur quite frequently, and there can also be reflex bradycardia during a hypertensive crisis, as well as a normal or even reduced heart rate (14).

In pheochromocytoma, there is often hyperglycemia linked to carbohydrate intolerance, accompanied by hypercalcemia and the presence of lactic acid in the peripheral blood (15). If the patient has severe hypertension, proteinuria may also occur (16). Many diagnostic tests, such as the measurement of plasma catecholamines or 24-hour urine, can only be performed in hospital or on an outpatient basis (17). Therefore, unfortunately, the diagnosis may not be immediately available.

Another diagnostic index is represented by the dosage of norepinephrine and epinephrine in the urine over 24 hours (18). After having ascertained the hypersecretion of catecholamines, procedures can be performed to search for the location of the tumor through magnetic resonance imaging (MRI) and computerized tomography (CT) of the abdomen (19,20). Scintigraphy can be useful in some cases, but not ultrasound (21).

However, the triad represented by headache and sweating that is accompanied by hypertension is a diagnostic index for pheochromocytoma (22). Other symptoms include panic attacks, hyperthyroidism, thyrotoxicosis, ischemic heart disease, amphetamine intoxication, menopause, and migraines (23). The presence of these diagnostic signs, especially combined with hypertension, is evidence to support the diagnosis of pheochromocytoma (24) (Table I). Hypertension is the most important diagnostic sign as it is almost always present. The patient with severe hypertension must be treated pharmacologically and the blood pressure must not be lowered quickly, bringing the diastolic pressure below 120 mm/Hg (25).

Table I. *Some of the clinical exams and patient symptoms which are utilized for the diagnosis of pheochromocytoma.*

Clinical exams:	Complete blood count, screening for electrolytes, urea nitrogen, creatinine, blood reticular acid measurements.
History of symptoms:	Anxiety, headache, sweating, heart palpitations, abdominal pain, chest pain, and weight loss.

Today, diagnostic imaging has significantly improved, and the localization of lesions is more precise, with better specificity and sensitivity. In addition, nuclear medicine provides a more powerful imaging modality than planar imaging, providing important information about the location and quality of lesions (26).

CONCLUSIONS

Pheochromocytoma is a tumor that forms in the adrenal medulla and is hormonally active. The tumor most often presents with hypertension, which is the most important diagnostic parameter. Patients suffering from this disorder may present paroxysmal attacks with headache, sweating, palpitations, anxiety, chest pain, and more rarely, nausea, vomiting, tremor, weakness, weight loss, and reduced breathing. The diagnosis of pheochromocytoma is difficult and often confused with more common pathologies such as preeclampsia, eclampsia, and thrombocytopenia. However, clinical exams such as MRI, CT, and scintigraphy allow for localization of the tumor.

Conflict of interest

The authors declare that they have no conflict of interest.

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