SECONDARY HYPERTENSION TO PHEOCHROMOCYTOMA: A CASE REPORT

HIPERTENSÃO SECUNDÁRIA AO FEOCROMOCITOMA: RELATO DE CASO

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ABSTRACT

Pheochromocytomas (PCC) are tumors that originate from chromaffin cells, which produce catecholamines, and are typically located in the adrenal glands. This rare condition has an incidence of 0.8 per 100,000 person-years, with a higher prevalence in women aged between 40 and 59 years. One of the main clinical manifestations is hypertension, classified as secondary hypertension due to PCC. We reported the case of a 53-year-old woman who sought medical care at a private cardiology clinic for a routine consultation and preoperative assessment for tumor excision in the right adrenal gland. She had previously been diagnosed with secondary arterial hypertension due to PCC. This type of tumor causes high blood pressure that requires pharmacological adrenal blockade until tumor excision. Normalization of blood pressure is expected after the procedure (i.e., adrenergic stimulus), with the patient returning to a normotensive state.

Keywords: Hypertension; Pheochromocytoma; Adrenal Glands.

RESUMO

Os feocromocitomas são tumores de células cromafins produtoras de catecolaminas, que podem ocorrer nas glândulas adrenais. É considerado raro, atingindo 0,8 por 100.000 pessoas/ano, com maior prevalência em mulheres entre 40 a 59 anos. Um dos problemas acarretados *é a hipertensão, sendo classificada como hipertensão secundária ao feocromocitoma. Relatamos o caso de* uma mulher de 53 anos, que procurou auxílio médico em uma clínica particular especializada em cardiologia, para consulta de rotina e avaliação pré-operatória para fins de exérese de tumoração em glândula adrenal direita. A mesma relata ter sido diagnosticada com hipertensão arterial secundáriaaofeocromocitoma. Ofeocromocitoma é uma causa de hipertensão secundária que resulta no aumento dos níveis tensionais, sendo necessário o bloqueio adrenérgico, até que seja feita a excisão do tumor, quando se espera que haja a normalização dos níveis pressóricos, à medida que o estímulo adrenérgico é superado com a retirada da neoplasia, voltando o paciente ao seu estado de normotensão.

Palavras-chave: Hipertensão; Feocromocitoma; Glândulas Suprarrenais.

INTRODUCTION

Systemic arterial hypertension (SAH) is a multifactorial clinical condition with high prevalence, defined by sustained blood pressure (BP) levels of ≥ 140/90 mmHg[1]. This condition is a major risk factor for other comorbidities, including acute myocardial infarction and stroke.

The prevalence of SAH in Brazil is variable, ranging from 2.5% to 30.9%; the prevalence increases with advancing age[2,3].

SAH can be primary or secondary. Primary SAH accounts for about 95.0% of cases and is associated with risk factors, such as genetic predisposition, smoking, obesity, and black skin color[4]. In these cases, the condition is considered chronic and incurable, with management focused on BP control.

Secondary SAH is less common, accounting for about 5.0% of cases. It occurs when patients with previously normal BP levels develop SAH following an underlying disease, including chronic kidney disease, obstructive sleep apnea or hypopnea syndrome, primary hyperparathyroidism, and pheochromocytoma (PCC). In contrast to primary SAH, secondary SAH can often be resolved with appropriate treatment of the disease, in addition to SAH treatment. However, prolonged exposure to elevated BP during secondary SAH promotes arterial stiffening, a risk factor for primary SAH.

PCC is a neuroendocrine tumor originating from chromaffin cells of the adrenal medulla. These tumors produce, store, metabolize, and secrete catecholamines[5]. They are rare, with an estimated prevalence of 0.1% to 0.2% among patients with SAH[6] across all ages; however, they are most prevalent between the ages of 40 and 59, and rarely after 60 years of age.

CASE REPORT

A 53-year-old woman (C.L.), white, married, teacher, attended a cardiologist for a routine consultation and preoperative evaluation for tumor excision in the right adrenal gland. She reported a diagnosis of SAH following clinical suspicion of PCC. The diagnosis was based on an abdominal and pelvic computed tomography scan with non-ionic contrast, which revealed an expansive lesion in the left adrenal gland measuring 3.2 x 2.8 cm, with subtle contrast enhancement during the imaging phases. During a consultation with her general surgeon, she exhibited a BP of 150/105 mmHg, and atenolol (100 mg orally) was prescribed once daily. Laboratory investigations revealed elevations of vanillylmandelic acid and urinary metanephrines. Physical examination at cardiology consultation showed a BP of 140/100 mmHg and a heart rate of 73 bpm. Then, she underwent home blood pressure monitoring (HBPM), which demonstrated normal values, and she was cleared for the proposed surgery. Histopathological findings of the surgical specimen revealed macroscopic alterations described as "irregular, elastic, yellowish tissue formation, weighing 38.18 g and measuring 5.5 x 3.5 x 3.0 cm. Sections disclosed a yellowish nodular lesion measuring 4.0 x 3.5 x 3.0 cm, centered by a cystic cavity of 3.0 x 2.8 cm filled with brownish fluid." Microscopy analysis reported "adrenal medullary hyperplasia origin composed of cells with abundant, basophilic, granular cytoplasm and small, clear nuclei, occasionally with visible nucleoli, arranged in nests surrounded by sustentacular cells and delicate fibrous septa. The foci of a fusiform cell pattern were observed. Two mitotic figures were identified in 10 high-growth fields, with tumor-free surgical margins". The final diagnosis was PCC with a pheochromocytoma of the adrenal gland scaled score (PASS) of 2, suggesting benign behavior (i.e., scores below 4)[7]. After the results, the patient returned to her cardiologist, who confirmed the diagnosis. Following the surgery, the patient returned to a cardiologic consultation, still using antihypertensive medication and presenting a BP of 110/70 mmHg and a heart rate of 70 bpm; she was instructed to discontinue medication gradually. After 30 days of medication discontinuity, a repeat HBPM revealed normal BP values. Another evaluation was conducted nine months later, confirming sustained normotension without the need for antihypertensive medication.

COMMENT

PCCs are neuroendocrine tumors arising from chromaffin cells of the adrenal medulla. They manifest a wide range of signs and symptoms, resulting in a heterogeneous and complex clinical presentation, with a high risk of cardiovascular morbidity and mortality[8]. Among these manifestations, SAH stands out as a highly prevalent and multifactorial condition.

PCC-related SAH is classified as secondary SAH. Initial control of secondary SAH may be achieved with pharmacotherapy, as demonstrated in this case with atenolol. Nevertheless, BP levels normalize with the removal of the PCC, resolving secondary AH.

In this report, the patient presents a case of secondary SAH, with BP normalized after tumor excision and histopathological confirmation of PCC. Additionally, the tumor was benign based on the PASS. Following surgery, her BP stabilized at 110/70 mmHg, and antihypertensive therapy was discontinued. The resolution of hypertension was confirmed in the 90-day follow-up assessment (including HBPM), supporting the effectiveness of the surgery.

Although the patient no longer has SAH, prolonged exposure to elevated BP levels may have induced arterial stiffness, increasing the long-term risk of developing primary SAH.

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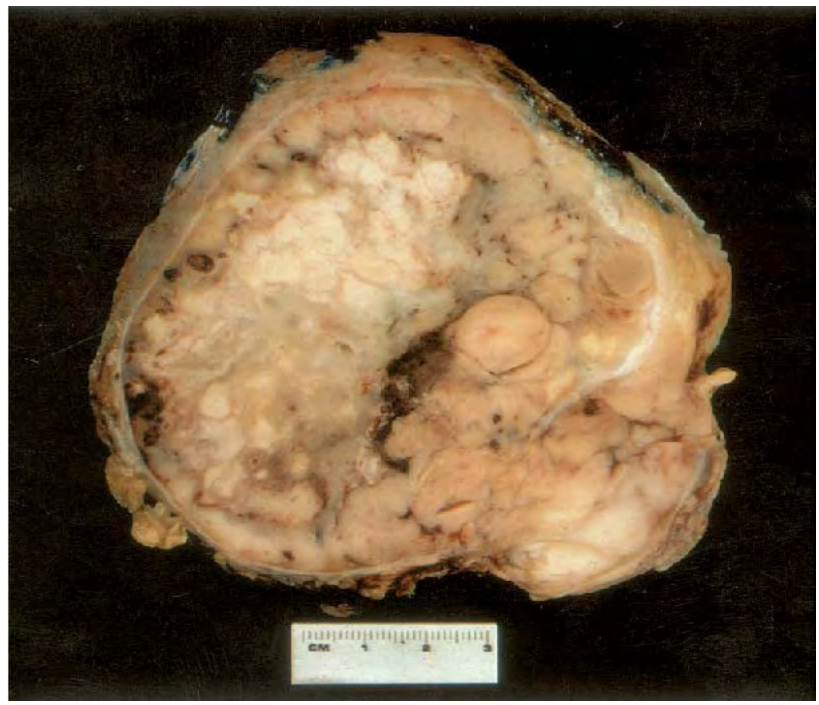

Figure 1. Macroscopic appearance of a pheochromocytoma tumor[7].

Pheochromocytoma of the Adrenal Gland Scale	
Microscopic Feature	Score
Vascular invasion	1
Capsular invasion	1
Periadrenal adipose tissue invasion	2
Cell nests of large proportions or Diffuse growth	2
Focal necrosis or confluent	2
High cellularity	2
Cellular Monotony	2
Mitotic figures > than 3 in 10 fields of great increase	2
Atypical mitotic figures	2
Marked Nuclear Pleomorphism	1
Hyperchromasia	1

Table 2. Pheochromocytoma of the adrenal gland scale score (PASS) according to Thompson's (2002)[7].