

## Bilateral Pheochromocytoma- A Diagnostic Dilemma

J. Ait Si Abdessadeq<sup>1\*</sup>, K. Outaghyame<sup>1</sup>, Y. Bouktib<sup>1</sup>, A. El Hajjami<sup>1</sup>, B. Boutakioute<sup>1</sup>, M. Ouali Idrissi<sup>1</sup>, N. Cherif Idrissi<sup>1</sup>

<sup>1</sup>Radiology Department, Er-Razi Hospital, Mohamed VI University Hospital, Marrakech, Morocco

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\*Corresponding author: J. Ait Si Abdessadeq

Radiology Department, Er-Razi Hospital, Mohamed VI University Hospital, Marrakech, Morocco

### Abstract

### Case Report

Bilateral pheochromocytomas are rare and are most often associated with familial diseases. However, sporadic bilateral cases can also occur. Therefore, in any patient with bilateral adrenal masses, a bilateral pheochromocytoma should be suspected, and it is important to determine whether it is familial or sporadic, given the potential risk of tumor recurrence. In this regard, we report the case of a 42-year-old patient, followed for moderate heart failure, who was hospitalized for investigation of newly discovered hypertension. Urinary levels of methoxylated derivatives were very high. Abdominal CT scans revealed the presence of two bilateral adrenal masses. MIBG scintigraphy showed increased uptake in both adrenal glands, without any other localizations. Investigations for multiple endocrine neoplasia or phacomatosis were negative. Treatment consisted of bilateral adrenalectomy via laparoscopy. The patient's progress under hydrocortisone replacement therapy was favorable, with normalization of blood pressure.

**Keywords:** Bilateral pheochromocytoma, adrenal gland, Arterial Hypertension.

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## INTRODUCTION

Pheochromocytoma is a rare tumor that develops from chromaffin cells, most commonly in the adrenal medulla, producing an excess of catecholamines.

Adrenal involvement is most often unilateral. However, in 10% of cases, a bilateral localization is observed. This presents several challenges regarding its diagnosis, malignancy criteria, and genetic aspects, especially in the absence of family history. Indeed, a mutation can be found in 25% of cases, even in patients with an "apparently" sporadic pheochromocytoma. The prognostic implications of these forms for both the patient and their family depend on the characteristics of each genetic syndrome.

## OBSERVATION

we report the case of a 42-year-old patient, followed for moderate heart failure, who was

hospitalized for investigation of newly discovered hypertension. The association with headaches, sweating, and palpitations led to the diagnosis of pheochromocytoma

The CT showed large (>5cm), well defined heterogeneous lesions in the bilateral adrenal gland region.

**on the right side:** oval-shaped, with poly lobed contours, well-defined, of tissue density, heterogeneous, with peripheral and punctate microcalcifications, showing heterogeneous enhancement after contrast injection, delineating a large area of central necrosis. This mass exerts scalloping on the superior pole of the right kidney.

**On the left side:** adrenal nodular formation, oval-shaped, with regular contours, spontaneously isodense and heterogeneous, showing heterogeneous enhancement after contrast injection, containing cystic areas.

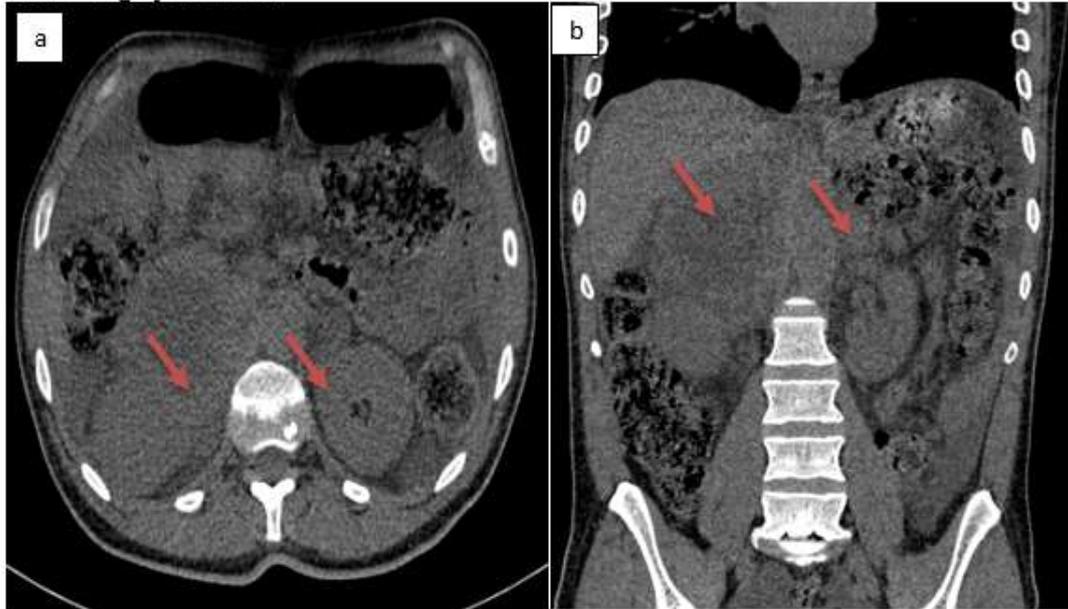


Figure 1 (a,b): Abdominal scan without contrast agent injection a: axial slice, b: coronal slice

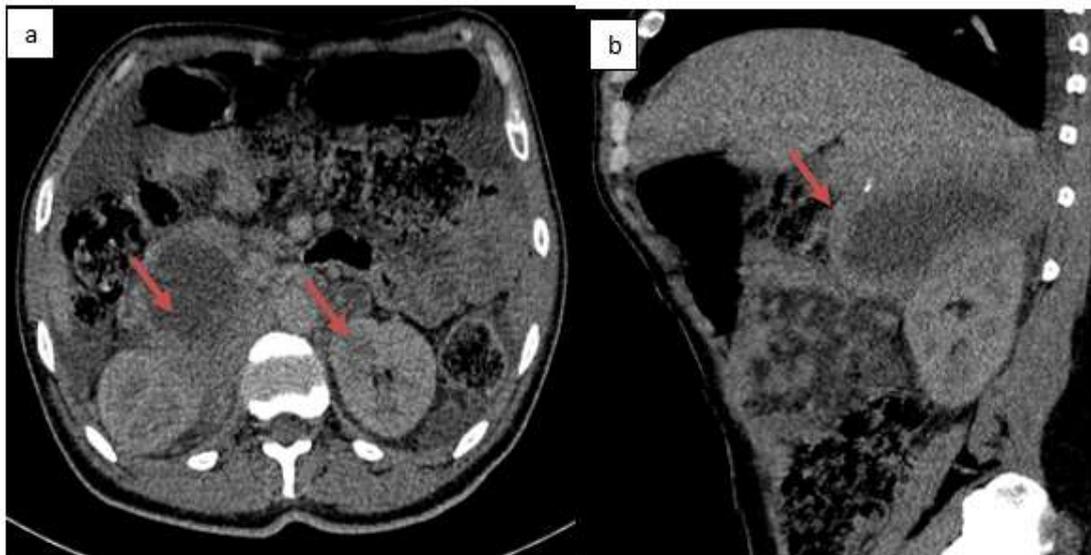


Figure 2 (a,b,c): Abdominal scan after contrast agent injection a: axial slice, b: right sagittal slice, c: left sagittal slice

## DISCUSSION

Pheochromocytomas are a type of paraganglioma originating from chromaffin cells. These tumors typically present with hypertension and/or cardiac arrhythmias. They can remain clinically silent when they grow larger. Imaging is essential for the localization, characterization, and staging of adrenal lesions, as well as for identifying extra-adrenal pheochromocytomas. Non-invasive diagnosis is crucial, as any physical contact with these tumors can trigger arrhythmias and malignant hypertension. Lab findings usually show elevated plasma metanephrines and increased catecholamine levels [1]. Most pheochromocytomas are located in the adrenal glands (adrenal pheochromocytoma), while some can be found

extra-adrenally. Diagnostic challenges arise due to the diverse imaging characteristics of this tumor.

Ultrasound imaging can show a range of appearances, from solid masses to mixed cystic and solid lesions, sometimes with necrotic or hemorrhagic areas [2]. In cases of acute hemorrhage within a pheochromocytoma, the mass may appear echogenic.

On computed tomography (CT), pheochromocytomas can be homogeneous or heterogeneous, solid or complex cystic masses with attenuation values greater than 10 HU. A few of them may show calcifications. They enhance significantly but may appear heterogeneous or show areas without enhancement due to cystic changes or necrosis.

Typically, they enhance more during the early venous phase than in the arterial phase, with contrast washout patterns similar to adrenal adenomas, with absolute and relative washout values greater than 60% and 40%, respectively [1, 3, 4]. On magnetic resonance imaging (MRI), pheochromocytomas typically appear hypointense on T1-weighted images and intensely hyperintense on T2-weighted images, often referred to as the "light bulb sign." These lesions show avid enhancement with gadolinium-based contrast agents, although the signal and enhancement patterns may vary depending on the presence of fat, hemorrhage, cystic changes, and necrosis [2].

I-123 meta-iodobenzylguanidine (MIBG) uptake in an adrenal nodule is strongly indicative of pheochromocytoma [2]. Positron emission tomography (PET) using dopamine is also highly sensitive for detecting extra-adrenal pheochromocytomas. Key differentiating factors between pheochromocytoma and adrenal carcinoma include well-defined, regular borders without evidence of adjacent infiltration, no lymph node involvement, and contrast washout rates greater than 60% and 40% for absolute and relative washout, respectively. Although the contrast washout pattern may resemble that of an adrenal adenoma, the larger size of the lesion, attenuation greater than 10 HU on plain CT, values over 125 HU during the early venous phase, heterogeneous contrast enhancement on CECT, and the absence of signal loss in out-of-phase MRI images, help rule out the possibility of an adrenal adenoma.

## CONCLUSION

Diagnosing bilateral pheochromocytoma requires confirming that both adrenal tumors are secretory in nature. MIBG scintigraphy is useful in confirming the neuroendocrine characteristics of the masses, helping to exclude the possibility of an incidentaloma of a different origin. It is essential to systematically investigate any underlying genetic disorder.

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