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(CASE REPORT)

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# Bilateral pheochromocytoma: A case report

M. Ben lafqih *, N. Hassan Aden, S. Rafi, G. El Mghari and N. El Ansari

*Department of Endocrinology, Diabetes, Metabolic Diseases and Nutrition, Mohammed VI University Hospital, Marrakech, Morocco.*

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## Abstract

Pheochromocytoma is a rare tumor developed at the expense of chromaffin cells, the adrenal involvement is most often unilateral. But, in 10% of cases, a bilateral localization is observed. We report the case of a 62-year-old patient, without personal or family pathological history, was hospitalized for exploration of recurrent pain in the right hypochondrium. On examination, the patient reported headaches, palpitations and sweating, on biological examination, urinary methoxylates were elevated, adrenal CT showed a large tissue mass, with fine calcifications and central necrosis, measuring 12*12*8.2cm with a washout <50%, the MEN 1 test was negative, the genetic study was not available, the diagnosis of sporadic right pheochromocytoma was the most likely, The patient underwent a right surrenalectomy with anatopathological examination in favor of pheochromocytoma, in the framework of follow-up a MIBG scan was done at 1 year post-op, it showed the presence of a focus of abnormal and isolated capture opposite the left arm of the left adrenal measuring 10.8*8.3 mm, in favor of a pheochromocytoma, the patient was operated: left adrenalectomy and the anatomopathological examination was in favor of pheochromocytoma. In these patients with bilateral pheochromocytoma, an underlying genetic disorder can be diagnosed in 80% of cases. It is most often a multiple endocrine neoplasia type 2, apart from the genetic aspect of the disease, the discovery of bilateral involvement requires confirmation of the secretory or functional character of the two adrenal masses in view of the possibility of the association of a pheochromocytoma and an incidentaloma of another nature.

**Keywords:** Bilateral pheochromocytoma; MEN1; Surrenalectomy; MIBG; Incidentaloma

## 1. Introduction

Pheochromocytoma is a rare tumor developed at the expense of chromaffin cells, most often medullary adrenal producing an excess of catecholamines. The adrenal involvement is most often unilateral. However, in 10% of cases, a bilateral location is observed. [1] It still poses several problems relating to its diagnosis, the criteria of malignancy and genetic aspects, especially in the absence of a family history. Indeed, a mutation can be found in 25% of cases, even in patients with an "apparently" sporadic pheochromocytoma [2].

## 2. Case presentation

Patient aged 62 years, without any pathological history, without any notable personal or family pathological history, was hospitalized for exploration of recurrent right hypochondrial pain. On examination, the patient reported headaches, palpitations and sweating. The physical examination was unremarkable. On biology, urinary methoxylates were elevated: Metanephrine 25 times normal, Normetanephrine 16 times normal. Adrenal CT showed a large spontaneously hypodense right adrenal tissue mass, with fine calcifications and central necrosis, measuring 12*12*8.2cm with a

* Corresponding author: maryame ben lafqih, Department of Endocrinology, Diabetes, Metabolic Diseases and Nutrition, Mohammed VI University Hospital, Marrakech,Morocco.

washout <50%. The diagnosis of right pheochromocytoma was retained. The genetic study was not available, the investigation for multiple endocrine neoplasia was negative, a sporadic form was the most likely given the absence of a specific family history and the negative MEN 1work-up. The patient underwent a right adrenalectomy by laparoscopy. The anatomopathology of the surgical specimen showed a morphological aspect compatible with a pheochromocytoma with a PASS score of 4. As part of the follow-up, a MIBG scan was performed at 1 year postoperatively, which showed an abnormal and isolated focus of uptake on the left arm of the left adrenal gland measuring 10.8*8.3 mm, in favor of a pheochromocytoma. In addition to this functional finding, the patient reported no Menard's triad, no flush syndrome, no bone pain, no motor diarrhea, and no signs of dysthyroidism. Methoxylated derivatives were normal and adrenal CT was unremarkable. The patient was taken back, the procedure consisted of a left adrenalectomy. The anatomopathological results were in favor of a pheochromocytoma and the immunohistochemical study showed a PASS score of 4 with expression of anti PS 100 AC, anti chromogranin AC, anti synaptophysin AC and a Ki 67 of 3%. The postoperative course was simple. The evolution was favourable with 30mg/d of hydrocortisone.

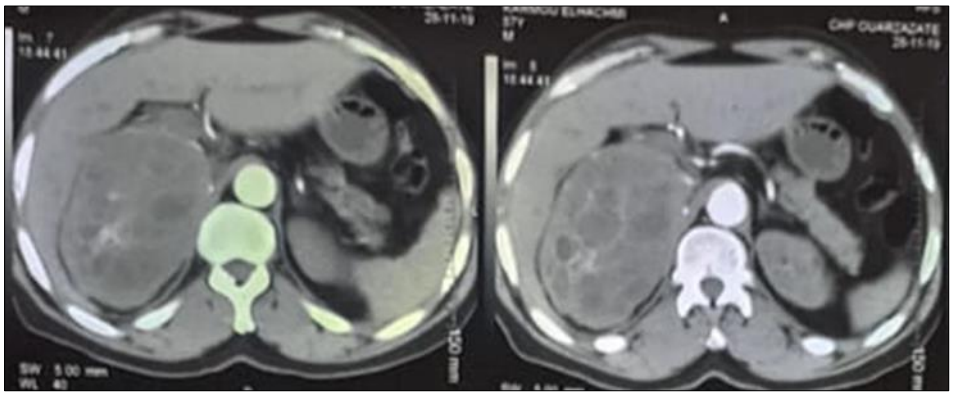
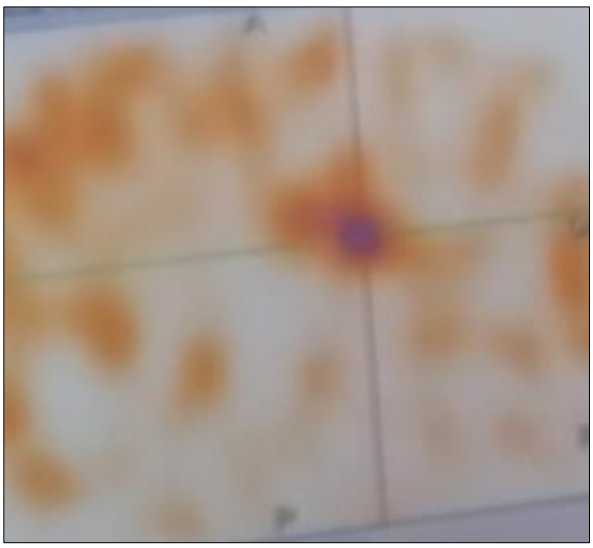


**Figure** Spontaneously hypodense right adrenal tissue mass, with fine calcifications and central necrosis, measuring 12*12*8.2cm



**MIBG scan:** An abnormal and isolated focus of uptake on the left arm of the left adrenal gland measuring 10.8*8.3 mm

## 3. Discussion

Pheochromocytoma is a neuroendocrine tumor that most commonly develops in the adrenal medulla. Pheochromocytoma is most often sporadic and unilateral. However, in 10% of cases it can be bilateral [1-4]. In these patients, an underlying genetic disorder can be diagnosed in 80% of cases [2]. This is most often multiple endocrine neoplasia type 2. The discovery of hereditary disease has great diagnostic and prognostic value for the patient and his or her family, as the probability of recurrence in familial pheochromocytomas is 13 times higher than in sporadic

pheochromocytomas [5]. Apart from the genetic aspect of the disease, the discovery of bilateral involvement requires confirmation of the secretory nature of both adrenal masses. Indeed, the existence of a typical clinico-biological picture of pheochromocytoma with bilateral adrenal nodules can evoke either a bilateral pheochromocytoma or the association of a pheochromocytoma and an incidentaloma of another nature. In this case, MIBG scintigraphy was of great interest. It allowed us to confirm the neuroendocrine character of the contralateral nodule and it guided us in the therapeutic decision [6]. Bilateral adrenalectomy is the treatment of choice for bilateral pheochromocytoma. As in our patient, replacement therapy is recommended for life to avoid adrenal insufficiency [7] with regular monitoring to detect other disorders in the context of MEN 1 that may appear.

## 4. Conclusion

Bilateral pheochromocytoma remains rare, the challenge is to differentiate a bilateral pheochromocytoma from a unilateral pheochromocytoma associated with an incidentaloma. MIBG scintigraphy is the best way to demonstrate the neuroendocrine character of both masses. The search for an underlying genetic disorder must be systematic and the surveillance must be regular.

## Compliance with ethical standards

### *Acknowledgments*

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### *Disclosure of conflict of interest*

I declare no conflict of interest.

### *Statement of ethical approval*

Informed consent was obtained from all individual participants included in the study.

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