

Extrasurrenal Pheochromocytoma: A Case Report and Review of the Literature

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Abstract

Introduction: Pheochromocytoma is a rare cause of endocrine hypertension. We report the case of an extra-adrenal pheochromocytoma discovered in the setting of an abdominal mass. **Observation:** The patient was 25-year-old and presented with headaches, excessive sweating and palpitations, followed by a sensation of abdominal weightlessness and diastolic arterial hypertension. Abdominal computed tomography revealed a retroperitoneal, right para-renal extra-adrenal mass. In biology, an increase in urinary methoxylated derivatives at the expense of normetanephrine. **Conclusion:** Pheochromocytoma is a rare disease, diagnosed by measuring urinary or plasma methoxylated derivatives. Conventional or nuclear imaging allows topographic diagnosis. Genetic studies helps to identify other tumors.

Keywords

Pheochromocytoma, Arterial Hypertension, Methoxylated Derivatives, Genetics

1. Introduction

Pheochromocytoma is a rare cause of secondary arterial hypertension.

Pheochromocytoma and paraganglioma are neuroendocrine tumors developed at the expense of chromaffin cells, capable of secreting excess catecholamines [1]. The prevalence of pheochromocytoma among cases of adrenal incidentaloma is approximately 5% [2]; the prevalence of paraganglioma in the

hypertensive population varies from 0.1 to 0.6% [3].

Pheochromocytoma is a rare condition in sub-Saharan Africa, due to limited health resources, with laboratory and imaging deficiencies causing our patients to be underdiagnosed [4].

Optimal management of these tumors requires multidisciplinary cooperation with a view to surgical excision.

We report a case of extra-adrenal pheochromocytoma in a 28-year-old young lady who consulted for a sensation of abdominal mass.

2. Medical Observation

A 25-year-old patient presenting, for about 6 months intermittently, headaches, palpitations and hyperhidrosis occurring on exertion and yielding spontaneously, is associated with a sensation of abdominal mass subject to abdominal ultrasound: retroperitoneal mass. She does not report renal colic or bone pain. Admitted to urology and then to endocrinology for exploration of this mass. On clinical examination, we noted a good general condition, diastolic arterial hypertension in particular at 130/110 mmHg in the left arm, 130/100 mmHg in the right arm, no hypotension orthostatic, a tachycardia at 100 beats/min; auscultation of the heart and vascular axes is unremarkable. The thyroid is not palpable, no nodules or cervical lymphadenopathy. Palpation does not reveal bone pain. Furthermore, there was no facio-truncal obesity or hirsutism. On imaging, in particular an abdominal computed tomography scan (**Figure 1**): Presence at the retroperitoneal, right pararenal level of a rounded tissue mass with regular contours, heterogeneous, enhanced after injection of contrast medium, measuring 50 × 61 mm. Douglas fir effusion blade compresses the inferior vena cava posteriorly and internally, which remains permeable.

It is in contact with the right kidney and the iliac PSOAS without signs of invasion, no deep adenopathy. It was a space mass whose epicenter is not at the level of the adrenal gland. The extension of the computed tomography at the level of the thoracic region was normal (**Figure 2**). On biology, the dosage of urinary methoxylated derivatives for 24 hours, with normal urinary creatinine,

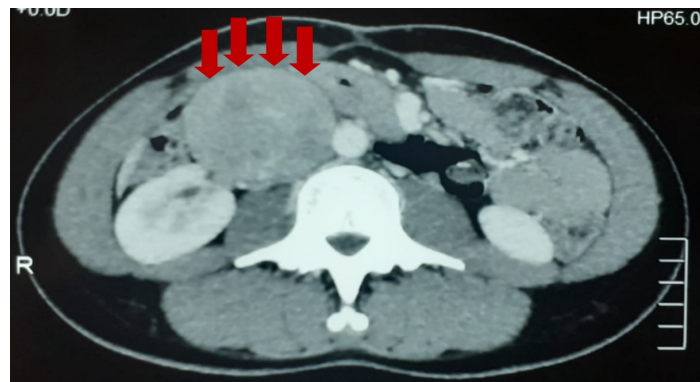


Figure 1. Abdomino-pelvic CT: cross section showing the 50 × 61 mm tumor (red arrows).

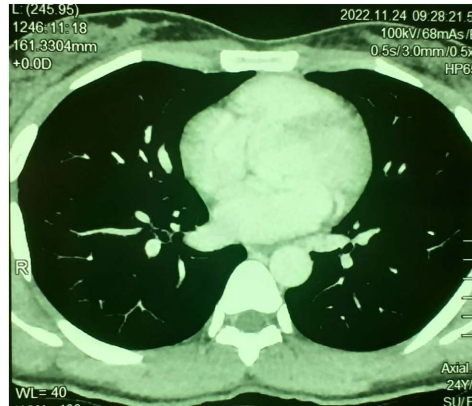


Figure 2. Chest CT: absence of tumor lesions.

reveals an increase in normetanephrine (4.6 times the upper value of the norm): 9.66 umol/L N: 0.40 to 2, 10; metanephrine was normal: 0.40 umol/L N: 0.20 to 1.00 (**Table 1**). The work-up for multiple endocrine neoplasia type 2, in particular a corrected calcemia at 89 mg/L, calcitoninemia 8 pg/mL and parathormoneemia at 25 ng/L were normal (**Table 2**). Trans-thoracic echocardiography revealed left ventricular hypertrophy preserved ventricular fraction at 69% (**Figure 3**). Genetic study to search genetic forms, including mutations SDHB, SDHC, SDHD, the RET proto-oncogene and of the VHL gene, could not be carried out in our work context. The optimal therapeutic strategy was discussed in a multidisciplinary consultation meeting including endocrinologists, cardiologists, anesthesiologists, urologists and visceral surgeons. Medical treatment based on alpha-blockers was initiated: prazosin 2 mg per daily orally and propranolol 80 mg per day in tablets for tachycardia. The surgical excision of the mass will be done in a foreign country. Our patient was evacuated to a foreign country because of the requirements of pheochromocytoma surgery, in particular the perioperative cardiovascular risk.

3. Discussion

Pheochromocytoma is a rare disease, involving hypersecretion of catecholamines. Pheochromocytoma may be adrenal localization (77%) or paraganglioma, of extra-adrenal localization (23%) located along the vascular-nervous axes mainly of abdominal (para-aortic), rarely thoracic, pelvic or cervical [5].

In our case, the location of the mass is extra-adrenal, abdominal at the retroperitoneal level, right pararenal.

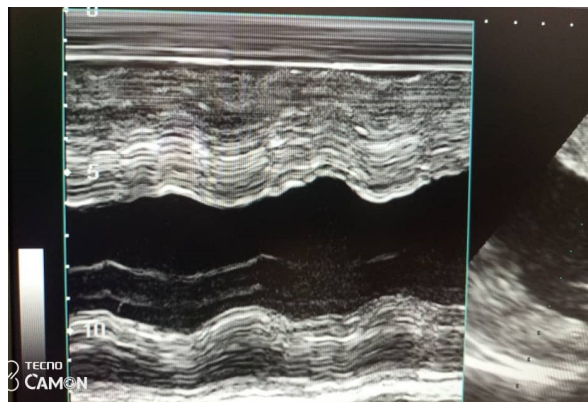
Pheochromocytoma is characterized by a great variability of biological activity and therefore in its clinical manifestations. Arterial hypertension is the most frequent sign, and can be permanent (50% - 60% of cases) or paroxysmal (35%), usually associated with sinus tachycardia. This arterial hypertension is secondary to the increase in vascular resistance, due to the action of catecholamines on the vascular alpha-adrenergic receptors. Orthostatic arterial hypotension may be observed by sympathetic vascular dysregulation [6]. Paroxysmal manifestations

Table 1. The results of the determination of urinary methoxylated derivatives.

	Patient values	Standards
Normetanephrine	9.66 umol/L	N: 0.40 to 2.10
	1.09 mg/L	N: 0.07 to 0.38
Metanephrine	0.40 umol/L	N: 0.20 to 1.00
	0.08 mg/L	N: 0.04 to 0.20

Table 2. Results of assessment of multiple endocrine neoplasia type 2.

	Patient values	Standards
Corrected serum calcium	89 mg/L	N: 84 - 105
calcitoninemia	8 µg/mL	N: 0 - 10
Parathormonemia 1 - 84	25 ng/L	N: 10 - 65

**Figure 3.** Echocardiography: left ventricular hypertrophy in long axis parasternal section. 69% LVEF TM Mode.

may be associated in particular with headaches (60% - 90%), palpitations (50% - 70%) and sweating (55% - 75%) [6], making up the Ménard's triad, which has a specificity of 94% and a sensitivity of 91% to evoke the diagnosis of a pheochromocytoma [7]. In our case, our patient, did not have orthostatic hypotension, presented permanent diastolic arterial hypertension, tachycardia associated with Ménard's triad occurring paroxysmally.

The biological diagnosis of hypersecretion of catecholamines is based on the measurement of plasma or urinary metanephrine because they have a sensitivity and specificity greater than 90%. The diagnosis is highly probable when their concentration is more than 3 times the upper limit of the norm [8] [9].

In our case, the 24-hour urinary metanephrine assay was performed. An increase of 4.6 times the upper value of the norm was noted at the expense of normetanephrine, consistent with an extra-adrenal pheochromocytoma.

In case of biological suspicion, the topographic diagnosis is based on the realization of a thoraco-abdomino-pelvic CT scan [10], which, in our case, at the retroperitoneal level, right pararenal of a rounded tissue mass of regular con-

tours, heterogeneous, enhanced after injection of contrast product, measuring 50 × 61 mm, whose epicenter is not at the adrenal level. Nuclear imaging may be necessary to look for multifocal involvement or secondary localizations. Genetic study in the case of pheochromocytoma or paraganglioma is essential, even in cases of sporadic pheochromocytoma, somatic mutations in the same predisposition genes can be demonstrated [11]. Several predisposition genes are associated with pheochromocytoma or paraganglioma: the RET gene; the VHL gene; the NF1 gene; SDHB, SDHD, SDHC, SDHA genes [12]. The genetic survey makes it possible to adapt the monitoring of the patient in relation to the mutation found, to detect the other tumors associated with this mutation. In our work context, genetic studies and nuclear imaging are not available. However, we carried out the phosphocalcic balance, the dosage of parathyroid hormone and calcitonin in the context of the search for multiple endocrine neoplasia type 2.

Medical treatment in preparation for surgery is essential. This medical treatment lasts 7 to 15 days before surgery and is based on rehydration and control of blood pressure. Alpha-blockers are used first line, in particular prazosin [13]. In our case, prazosin was instituted as a medical treatment.

Curative treatment remains surgery, adrenalectomy or lumpectomy. The main risk is an adrenergic discharge which can lead to a hypertensive crisis and/or cardiac arrhythmia, in particular during induction of anesthesia, intubation or manipulation of the tumor.

The multidisciplinary consultation meeting required surgery for our patient in a better-equipped work environment. At the end of this surgery, the histo-pathological study will be carried out. Postoperatively, a determination of plasma and/or urinary metanephrine will be carried out 2 to 6 weeks after surgery. According to international recommendations, in the event of normalization, follow-up will be annual by consultation and measurement of plasma and/or urinary metanephrine [14].

4. Conclusion

Pheochromocytoma is a rare cause of secondary hypertension of endocrine origin. Its positive diagnosis requires the dosage of urinary methoxylated derivatives for 24 hours, and its adequate management requires the involvement of a multidisciplinary team, the possibility of carrying out a genetic investigation and an efficient technical platform.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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