SUNTEXT REVIEWS

SunText Review of Case Reports & Images
ISSN: 2766-4589

Open Access
Research Article
Volume 6:4

Laparoscopic Treatment of a Secreting Retroperitoneal Extra-Adrenal Pheochromocytoma (Paraganglioma): A Case Report and Review of the Literature

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Received date: 20 Septmeber 2025; **Accepted date:** 15 October 2025; **Published date:** 25 October 2025

Citation: Habarek M, Yahiaoui T (2025) Laparoscopic Treatment of a Secreting Retroperitoneal Extra-Adrenal Pheochromocytoma (Paraganglioma): A Case Report and Review of the Literature. SunText Rev Case Rep Image 6(4): 164.

DOI: https://doi.org/10.51737/2766-4589.2025.164

Abstract

Background: Paraganglioma is an endocrine tumor developed at the expense of extra-adrenal chromaffin cells. Retroperitoneal forms are less frequent than other locations. Paraganglioma has the same embryological origin and the same histological structure as pheochromocytoma. Their clinical presentation is similar, and depends essentially on the hormonal secretion of the tumor. The positive diagnosis of paraganglioma requires plasma and urine hormonal assays. Imaging and isotopic explorations are essential before surgery. Surgery is the only curative therapeutic option. It is associated with the prevention and monitoring of hemodynamic and cardiovascular disorders, which are frequently observed. The prognosis depends on the metastatic nature of the tumor and the presence of postoperative tumor residue. A genetic investigation is systematically proposed, especially since there is a correlation between tumor aggressiveness and the type of genetic mutation. The aim of our study is to report a new, rare observation of secreting retroperitoneal paraganglioma treated laparoscopically.

Material and Methods: The authors report a case of secreting retroperitoneal paraganglioma, operated in the general surgery department of the TiziOuzou University Hospital, between 2016 and 2024. It was a 39-year-old woman. The clinical data are not specific and the discovery may be fortuitous. The laparoscopic approach was performed.

Results: The patient had abdominal pain and treatment-resistant hypertension with palpitations, associated with a suspicious retroperitoneal mass on imaging and an increase in urinary methoxylated derivatives preoperatively. The mean follow-up was 31 months. Hypertension and palpitations had regressed after surgery. No local or secondary recurrence on control thoracoabdominal CT scan.

Conclusion: Retroperitoneal paraganglioma is rarely observed. The clinical symptomatology is not specific, and may be similar to that of pheochromocytoma. Abdominal CT and MRI, associated with MIBG scintigraphy are highly suggestive. The management of paraganglioma must be multidisciplinary but only surgical treatment is curative and prevents recurrences.

Keywords: Paraganglioma; Pheochromocytoma; Retroperitoneal tumor; Anesthesia; Surgery

Introduction

Paragangliomas (PG), or extra-adrenal pheochromocytomas, are rare neuroendocrine tumors developed at the expense of the parasympathetic nervous system (neuroectodermal cells, or paraganglionic tissue) [1]. They are defined as extra-adrenal chromaffin tumors and represent approximately 1/5th of chromaffin tumors. The adrenal site is usual (90%), the extra-adrenal location is rare representing 10% of paragangliomas with

an incidence rate of 2-8 cases per million people/year [2]. They have a variable topography and the functional retroperitoneal form represents 2% of cases [3]. The retroperitoneal and subdiaphragmatic forms would be less frequent than the other locations (head and neck) [4]. The retroperitoneal paraganglioma is an uncommon situation [4-7]. Their clinical diagnosis is difficult because they evolve slowly [8,9]. These tumors, when they are secreting, are characterized by excessive production of catecholamines [10-13]. The prevalence of these tumors varies between 0.2 and 0.6% of patients with hypertension [14]. It is important to diagnose and treat them because of a significant increase in cardiovascular morbidity and mortality associated with them [15].

Histology cannot formally rule out the malignancy of the lesion. Malignancy is defined by the presence of secondary localization in non-chromaffin tissue and represents approximately 10% of these tumors [15]. However, with the advancement of research and a better understanding of this disease over the last decade, it appears that the terms "benign" and "malignant" have a less and less important meaning, because all PG has a metastatic risk, and tend to be replaced by a risk stratification strategy [10,16]. This set of diseases represents a real challenge in terms of diagnosis and therapeutic management [17]. The malignant potential of these tumors requires the use of excisional surgery. The management of paragangliomas must be multidisciplinary but only surgical treatment is curative. We report the case of a 37-year-old female patient, treated for a secreting PG located in the left flank, who has intimate contact with the left kidney and its pedicle. From this clinical case, we will review the particularities of the management of this pathology from a diagnostic and therapeutic point of view.

Material and Methods

Patient I. K., 39 years old, with a family history of high blood pressure in the father, followed and treated, with deep vein thrombosis treated with Sintrom for more than 6 months. The clinical history of this woman began with abdominal pain in the left flank. This pain appears several times a day, at rest, sometimes accompanied by palpitations and dizziness. The clinical signs have been noted. Therapeutic methods and short- and long-term results were noted. The surgical indication and the choice of the type of surgical treatment, on the basis of the immediate results are also under discussion.

Results

The clinical examination revealed a normal BP of 130/80 mm Hg, a BMI of 21.05 kg/m2 for a weight of 58 kg and a height of 1.66 m. The existence of Menard's triad was noted, consisting of pulsatile headaches, tachycardia at 105 beats/minute and profuse sweating. Furthermore, there are no other functional signs of hormonal hypersecretion, nor clinical signs that could suggest a genetic disease such as neurofibromatosis type 1 (MEN 1), multiple endocrine neoplasia type 2A (MEN2a) and finally Von Hippel Lindau disease. The rest of the clinical examination is within the normal limits.

A biological assessment is carried out, plasma free metanephrines/normetanephrines: metanephrines = 0.53 nmol/l (N < 0.33 nmol/l), normetanephrines = 11.50 nmol/l (N < 1.07 nmol/l). A 24-hour urine collection for a 24-hour urinary methoxylated derivative rate, namely: normetadrenaline> 4950 nmol (N < 281 nmol, i.e. 17 times the norm), metadrenaline = 421 nmol (N < 159 nmol, i.e. 2.67 times the norm), methyldopamine = 282 nmol (N < 329 nmol). A biological assessment of associated diseases is performed, namely, the tyrocalcitonin level = 0.95 pg/ml (normal) for NEM 2a diseases and the parathyroid assessment: PTH = 38.88 pg/ml (N: 15-65pg/ml), calcemia: 90 mg/l (normal), phosphoremia: 3.1 mg/l (normal) for NEM 1 diseases. Additional biological assays had eliminated anunusual hormonal secretion. On abdominal CT, an intra-abdominal mass is observed opposite the left flank, retroperitoneal, extra-adrenal, which has intimate contact with the left renal cortex, its pedicle and the left ureter without invading them. It measures 58 mm transverse axis x 50 mm anteroposterior axis x 50 mm height with regular contours, relatively homogeneous and strongly enhanced after injection of contrast agent. There is no infiltration of neighbouring organs (Figure 1).

Abdominopelvic MRI shows an intra-abdominal, retroperitoneal, roughly oval mass measuring 57/47/37 mm in long axes, with a hyperintense T2 and hypointense T1 signal, strongly enhanced in a heterogeneous manner in the arterial phase after injection of gadolinium. It comes into intimate contact with the left kidney and its vascular pedicle. The left renal vein is discreetly compressed (Figure 2). MIBG scintigraphy reveals an aspect of a secreting neuroectodermal process at the level of the left flank (paraganglioma) without secondary localizations at a distance (Figure 3). The diagnosis of retroperitoneal, extra-adrenal, secreting paraganglioma was retained. Medical treatment with prazosin (alpha blocker) was introduced with a view to surgical treatment. Laparoscopic surgery with 3 trocars was performed ten days later (1 trocar of 10 mm diameter for the optic, a trocar of 5 mm diameter for grasping and another operating trocar of 12 mm) (Figure 4). Induction was done by intravenous propofol (3 mg/kg) and sufentanil (0.5 µg/kg), orotracheal intubation facilitated by rocuronium (0.6 mg/kg); maintenance was ensured by a continuous infusion of propofol and rocuronium. Perioperative monitoring was provided by capnography, SpO2, ECG, temperature, diuresis, monitoring of curarization, blood pressure and placement of a central venous access (PVC). Blood pressure

was less than or equal to 180 mm Hg and a central venous pressure of 8 to 10 cm of water.

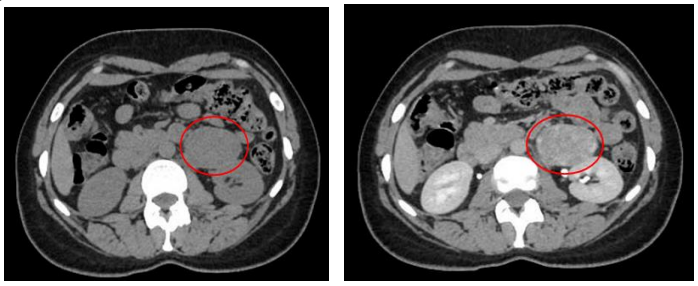

Figure 1: *CT scan showing the pseudoencapsulated retroperitoneal mass in contact with the left renal pedicle, measuring 58x50x50 mm and resting on the leftrenal vein.*

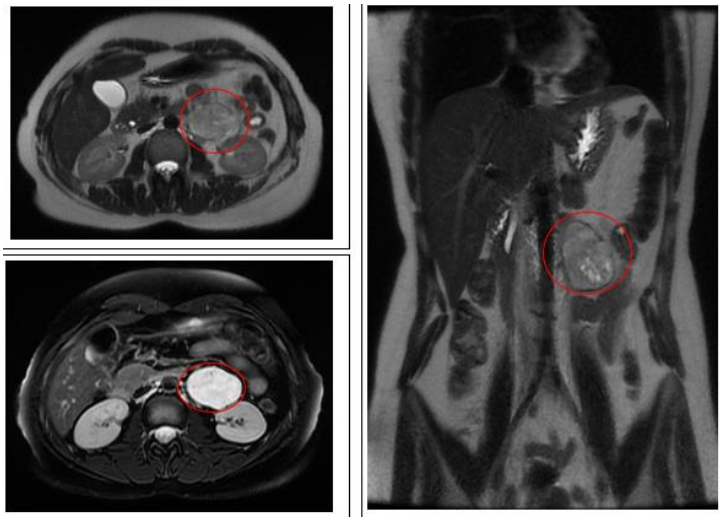

Figure 2: *MRI section highlighting the retroperitoneal mass measuring 57/47/37 mm with large axes of hyperintense T2 and hypointense T1 signal, strongly enhanced in a heterogeneous manner in the arterial phase after injection of gadolinium.*

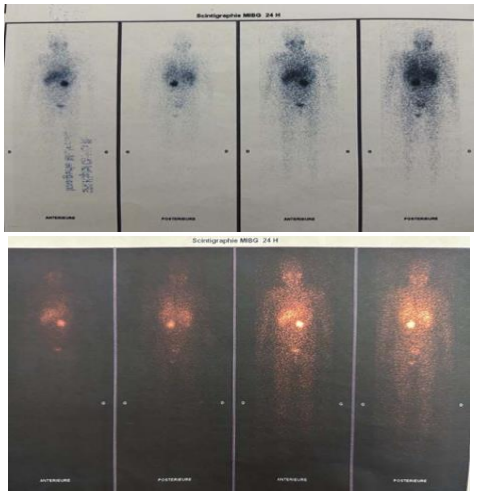

Figure 3: *MIBG tomoscintigraphy: very intense capture at the level of the tissue mass of the left flank (paraganglioma) without secondary locations at a distance.*

Figure 4: *Placement of the 3 abdominal trocars.*

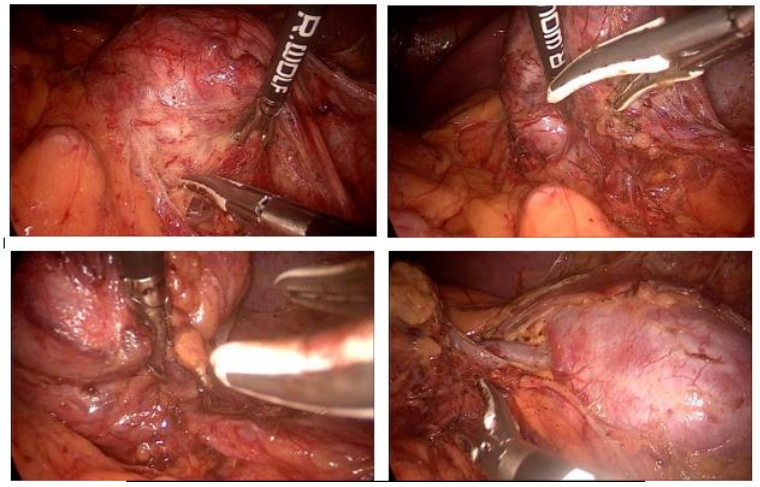

Figure 5: *Intraoperative images of the tumor and its relationships.*

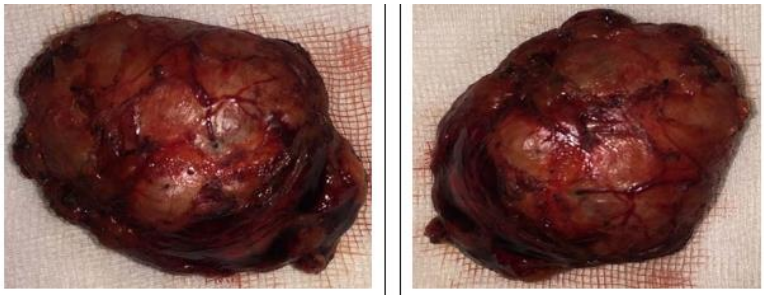

Figure 6: *Macroscopic aspect of the surgical specimen.*

Figure 7: *Fixed slices of the extra-adrenal tumor.*

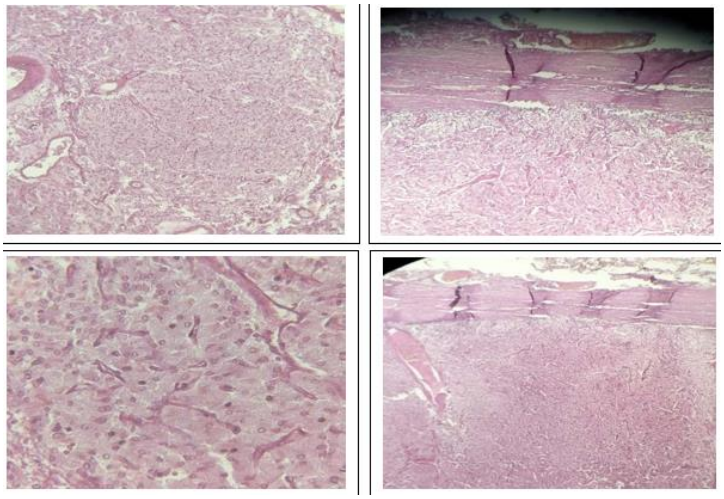

Figure 8: *Tumor proliferation of neuroendocrine nature with nested, trabecular or solid arrangement. Tumor cells are large, polygonal, widely vacuoles with abundant, fine and granular cytoplasm.*

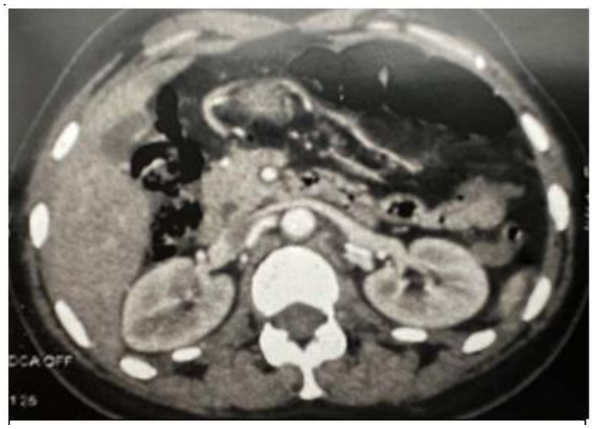

Figure 9: *Normal CT scan control after 30 months postoperatively.*

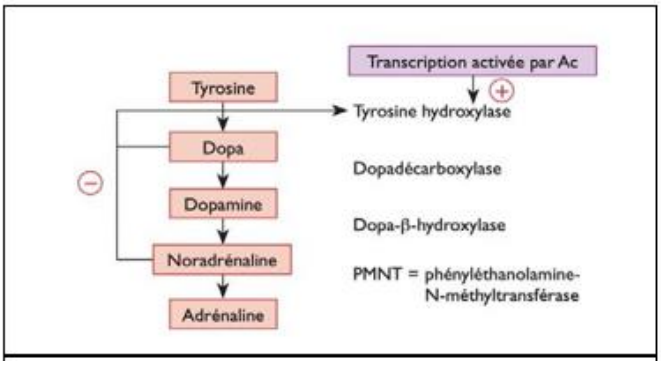

Figure 10: *Synthesis of catecholamines AC: acetylcholine.*

Perioperative exploration revealed after detachment of the left colon a retroperitoneal, extra- adrenal, left pararenal mass of 6 to 7 cm in diameter. The tumor is hypervascularized, firm, with regular contours, encapsulated. It adheres to neighbouring structures without invading them (the left ovarian vein and the left ureter). It compresses the left renal vein (Figure 5). The surgical technique included minimal mobilization of the tumor. The peritumoral pedicles were checked first. Intraoperatively, blood pressure peaks defined by a SBP > 160 mm Hg were treated with nicardipine boluses (2–4 mg) in order to bring the SBP between 120–160 mm Hg which was maintained by nicardipine at a dose varying between 3 and 6 mg/h by electric syringe. Four episodes of sinus tachycardia (heart rate [HR] greater than 100–120 beats per minute) were treated with intravenous boluses of esmolol 0.5 mg/kg with maintenance electric syringe at a dose of 50 µg/kg/min. Despite discontinuation of antihypertensives and good filling, introduction of noradrenaline at a dose of 0.5 mg/h was necessary after tumor resection. Complete excision of the mass was then performed without invasiveness. The surgical specimen was extracted in a plastic bag through the enlarged 12 mm trocar. The surgical procedure was completed without abdominal drainage. The duration of the procedure was 210 minutes. Postoperatively, during the first 24-48 hours, we established a monitoring sheet for blood pressure, heart rate and blood sugar levels in order to reduce postoperative complications, which are mainly hypotension, hypertension and hypoglycemia. The immediate postoperative course was simple. Urinary metanephrines and normetanephrines returned to normal postoperatively. On macroscopic examination, the mass weighed 285g and measured 75x65x60 mm. It was cystic and fleshy in appearance, with a thickened cyst wall (Figure 6).

On section, a mahogany yellow appearance was noted with areas of necrosis (Figure 7). Histological examination showed tumor proliferation made up of cord areas and clusters arranged around vascular structures creating a neuroendocrine architecture (Figure 8). This morphological and immunohistochemical analysis was in favour of a retroperitoneal paraganglioma with complete excision, without aggressive elements, without vascular emboli or evidence of extra-tumor extension. The patient was reviewed in consultation at one month, six months, one year, two years and 2 and a half years; her clinical examination, thebiological assessment: the urinary methoxylated derivatives of the 24 hours (normetadrinaline = 0.83 nmol, metadrinaline = 0.29 nmol) were normal. The control thoraco-abdomino-pelvic CT scan at two and a half years was without abnormality and did not show any recurrence (Figure 9). Due to the risk of recurrence persisting several years after complete surgical excision of the tumor, the patient will be followed up continuously on the clinical and biological levels with annual dosages of urinary metanephrines and normetanephrines supplemented by morphological and functional imaging in the event of an increase in metabolites.

Discussion

A distinction is made between catecholamine-secreting adrenal tumors calledpheochromocytomas and extra-adrenal tumors that may or may not secrete catecholamines, and called paragangliomas [18]. Paragangliomas are tumors that develop at

the expense of neuroectodermal cells of the autonomic nervous system. These cells, originating from the neural crest, migrate along the aorto-sympathetic axis to give rise to either sympathetic ganglion cells or paraganglionic cells by glandular differentiation. The latter usually involute around the age of three years, leaving only vestiges, with the exception of the adrenal medulla [19]. Different classifications have been proposed, taking into account the secreting or non-secreting nature of these tumors and their location. Functional paragangliomas, secreting catecholamines, chromaffin, were classically contrasted with non- functional tumors or chemodectomas, which are not chromaffin. This notion of chromaffinity should no longer be retained because there are non-chromaffin secreting tumors and non- chromaffin secreting tumors [20].

Currently, functional paragangliomas are distinguished from non-functional paragangliomas. Topographically, and in decreasing order of frequency, these tumors are subdiaphragmatic in 85% (para-aortic: 46%, Zuckerkandl's organ: 29% and bladder: 10%), thoracic in 12% and finally cervical in 3% [19]. Paragangliomas occur at an earlier age, 10 to 30 years but more frequent in young adults [20]. They are multifocal in 15 to 24% of cases [4]. They are much more often malignant than intra-adrenal pheochromocytomas approximately 40% against 10%. Malignant forms occur earlier than benign forms and are characterized by the occurrence of local invasion (lymphatic type) or distant invasion (invasion of the lung, bone, liver) in 30% of cases [9,20]. In approximately 30% of cases, paragangliomas are genetically determined [22]. The familial form of the disease is called hereditary paraganglioma. It is transmitted in an autosomal dominant manner with incomplete penetrance. In affected subjects, tumors appear early and are often multiple and/or recurrent from the outset [22]. The high frequency of familial forms justifies the systematic performance of a genetic investigation in any patient with a paraganglioma that is apparently sporadic [23]. Paragangliomas are most often isolated. They can be associated with other pathologies: Carney's triad (PG, gastric leiomyosarcoma and pulmonary chondroma), Multiple Endocrine Neoplasia (MEN) type 2, and PG associated with Von Hippel-Lindau disease (VHL) and neurofibromatosis type 1 (Recklinghausen disease) [24,25]. The immediate diagnostic and therapeutic implications due to the risk of tumor malignancy, the severity of complications and the curable nature of this condition require rapid management of functional paragangliomas in a specialized setting. The steps of this are based on the diagnosis of catecholaminergic hypersecretion, tumor localization and the search for possible metastases, therapeutic management by surgical excision and possible adjuvant treatment, genetic diagnosis and postoperative follow-up of the patient. In the present case, the lesions described on the initial imaging raised suspicion of a retroperitoneal paraganglioma, contrasting with the clinical picture of a patient with few symptoms at the time of diagnosis.

The clinical presentation of pheochromocytoma and intrathoracic or abdominal paragangliomas varies greatly. Symptoms and signs are often mediated by increased secretion of catecholamines (noradrenaline alone for intrathoracic or abdominal PGLs and noradrenaline/adrenaline for pheochromocytomas due to enzymatic activation of phenylethanolamine-N-methyltransferase by cortisol) (Figure 10) [26]. If the main symptom of pheochromocytoma or paraganglioma is high blood pressure, pheochromocytoma is the cause of only 0.5% of high blood pressure [27]. The triad of headache, palpitations, and profuse sweating is found in nearly 90% of cases [18,28] as is the case in our patient. The other symptoms are less suggestive: ascending abdominothoracic constrictive pain, anxiety, tremors, pallor, and digestive disorders. Retroperitoneal paraganglioma is a rare entity [27]. From a metabolic point of view, hyperglycemia, lactic acidosis, and weight loss are observed. Less frequently, patients have nausea, fever, and flushing due to the co-secretion of a multitude of peptides (vasointestinal peptide, substance P, interleukin-6) [14,26]. In 30% of cases, the clinical presenting sign is pain, depending on the location and size of the tumor, which may be intra-abdominal with a clinical presentation of lower back pain, renal colic or biliary colic. Finally, in 10 to 30% of cases, it is a fortuitous discovery on imaging performed in search of another complaint where pheochromocytomas represent approximately 5% of adrenal incidentalomas. Even more rarely, the diagnosis is established in the context of screening for a known germline mutation (Table 1) [14].

Table 1: *When to look for pheochromocytoma/paraganglioma?*

• Young patient with HBP without risk factors
• HBP with:
- Symptoms (weight loss)
- Orthostatic hypotension
- Unexplained shock
- Hyperglycemia with BMI 125 kg/m2
- Cardiomyopathy
- Resistance to treatment
• BP lability without other cause
• Family history of PHEO/PGL
• Medullary thyroid carcinoma, family history of medullary thyroid carcinoma, NF1, cutaneous neuromas
• When an adrenal incidentaloma is detected
• Shock or severe blood pressure response during a procedure

Table 2: *Conditions for collecting métanéphrines /normetanephrines.*

Patient preparation
- No taking of sympathomimetics (nicotine, amphetamines, adrenaline)
- No interfering treatment (labetalol, sotalol, methyldopa, SSRI, MAOI)
- No caffeine or theine in the 12 hours preceding
Plasma collection for free metanephrines
- Lying position, after 30 min of rest, butterfly pose
- Tube with heparin
24-hour urine collection
- Acidification of urine to PH 4

The first diagnostic step is to highlight the excess secretion of catecholamines. This is the case of our patient. The measurement of catecholamines, adrenaline and noradrenaline, is no longer recommended due to the lack of sensitivity and specificity [26]. The tests currently recommended are the measurement of free plasma metanephrines and normetanephrines or the measurement of fractionated urinary metanephrines and normetanephrines on a 24-hour urine collection. These are inactive metabolites of adrenaline and noradrenaline, produced by pheochromocytomas and paragangliomas [26]. These two tests have a good sensitivity > 95% with a slightly lower specificity at around 90-95%. However, they require prior preparation of the patient as well as specific conditions to avoid false positive results (Table 2). Falsenegatives are rare (three quarters of patients with PHEO/PGL have métanéphrines/normetanephrines elevated to 3 times the upper norm) but can be considered in the context of a very small tumor (< 1 cm), a necrotic tumor or in the absence of synthesis or metabolism of catecholamines by the tumor. It is also rare that an elevation of more than 3 times the norm of one or the other of the values is a false positive [26,29,30]. In patients with borderline results, these will have to be interpreted according to the clinic and on the basis of a second dosage but, in the absence of exclusion of any factor that could lead to false positives, the option is generally to continue the investigations with imaging.

After obtaining a positive biological result, imaging is indicated to localize the tumor. The two possible imaging methods are CT and MRI. The typical appearance of these tumors is that of a spherical or ovoid lesion, well demarcated, tissue, with a certain heterogeneity, necrotic areas and calcifications. The injection of contrast product depending on the type of imaging helps to characterize the lesion. Due to the predominantly intra-abdominal location of PHEO/PGL, an abdominal and pelvic CT/MRI is the imaging of choice [31]. MRI is recommended for patients with metastatic PGL and in patients who have a contraindication to exposure to radiation (pregnant women, children, etc.) [14]. So-called functional imaging, scintigraphic or by positron emission tomography, is of higher specificity for positive diagnosis, localization and assessment of tumor extension [32]. Scintigraphic imaging is recommended in the initial assessment for patients with large pheochromocytoma or PGL, conditions that increase the risk of possible metastatic dissemination. 123I-Meta-iodo-benzyl- guanidine (MIBG) scintigraphy remains an examination of choice in this indication. On the other hand, FDG-PET/CT would be superior to MIBG scintigraphy in the case of a known metastatic tumor [14]. In our observation, tomoscintigraphy showed very intense uptake at the level of the left pararenal tissue mass.

Treatment of paraganglioma requires multidisciplinary care. Surgery is the treatment of choice. First-line surgery is a laparoscopic approach for intra-adrenal pheochromocytomas. Regarding our patient who has a secreting retroperitoneal PGL, we opted for Coelioscopy because the topography and vascular relationships of the tumor allowed it. This approach reduces postoperative pain, the risk of bleeding, and the number of days of hospitalization compared to open surgery by laparotomy. The experience of the operators and anesthesiologists reduces the peroperative risk. Surgery remains the only curative treatment provided that it is complete [33]. It allows survival rates of 75 and 45% at five and ten years respectively [33]. Catecholamines are released during tumor manipulation with a withdrawal phenomenon upon removal of the surgical specimen, hence the interest in preparing these patients before the operation by progressively blocking the α and β receptors over one to two weeks to limit peroperative hypertensive episodes and restore blood volume. This preparation aims to control the permanent portion of arterial hypertension, correct hypovolemia and prevent rhythm disorders [34,35]. Good control of blood pressure reduces the risk of operative complications. In our observation, initial treatment with calcium channel blockers alone did not stabilize blood pressure, which led us to do a ten-day preparation combining a α and a β blocker. This preparation can be short (three to four days) if blood pressure is stable or longer (seven to ten days) in the event of blood pressure instability.

Anesthetic management is dominated by preoperative cardiac assessment because paraganglioma can cause a true adrenergic cardiomyopathy [28,34,36]. Left ventricular hypertrophy and sometimes obstructive cardiomyopathy can be observed, linked to the arterial hypertension developed by patients. Acute manifestations are possible, including episodes of pulmonary edema or pictures suggesting an acute coronary syndrome. The onset of left or global heart failure, even without any arterial hypertension, with a collapsed ejection fraction and global hypokinesia is possible [34]. Echocardiography is the most useful examination in this context [34,36]. It allows to assess the impact of catecholamine secretion and the effect of the preoperative drug preparation on these heart diseases [34,36]. It allows to set the optimum time for surgery. Premedication includes an anxiolytic and maintenance of the antihypertensive treatment established preoperatively. Perioperative monitoring, in addition to the standard elements, includes systematic measurement of blood pressure by blood. More or less invasive hemodynamic monitoring is useful to detect variations in right and left filling pressures during tumor manipulation or rapid volume expansion [37,38]. General anesthesia is based on non-histamine-releasing products. Deep anesthesia is essential. There is no specific recommendation for the choice of induction anesthetic agents, with the exception of the avoidance of certain products (ketamine, droperidol, metoclopramide, tricyclics, phenothiazines) due to indirect effects on catecholamine secretion [39]. Propofol is the

induction agent of choice. Thiopental has been used without risk. Etomidate is recommended as an appropriate agent in patients with unstable hemodynamic status [27]. Cisatracurium is the curare of choice for maintenance of hemodynamic stability. Fasciculations generated by succinylcholine can stimulate tumor secretion [27]. Maintenance may include high doses of sufentanil and inhalation of sevoflurane, chosen for its rapid kinetics. Desflurane is not the agent of choice due to sympathetic stimulation. Blood volume is continuously optimized.

Hypertensive surges triggered by intubation and tumor manipulation, arrhythmias and hypotension after tumor resection are the main events that can occur intraoperatively [34,39]. In our observation, these hypertensive surges and arrhythmias are managed by the administration of calcium channel blockers using an electric syringe and β blockers (Esmolol). Arterial hypotension, or even collapse after tumor removal, especially if it is a dopamine secretor, will benefit from early discontinuation of vasodilators and β-blockers as soon as the vein draining the tumor is ligated, and optimization of filling associated with noradrenaline, best guided by hemodynamic collection [37]. Hemodynamic instability may persist for several days due to abrupt withdrawal of catecholamines. Hypoglycemia is detected systematically due to the derepression of pancreatic insulin secretion. Postoperative care of a few hours, most often in the post-intervention monitoring room, is sufficient. It consists of hemodynamic monitoring, detection of hypoglycemia and surgical complications. A potential for tumor malignancy may be suspected on pathological examination but never confirmed. Only the presence of metastases defines with certainty a malignant functional paraganglioma. Several histological scores (including the PASS score – Pheochromocytoma of the Adrenal gland Scaled Score [40] have been proposed to assess tumor malignancy, taking into account the invasion of adjacent structures, the cell proliferation index or the cytological profile. These scores help to identify tumors at risk of metastases, but none of them can discriminate with certainty the malignant nature of the tumor or specify the schedule for subsequent follow-up [32,41]. Patients must have biological monitoring at three months, then annually (metanephrines/normetanephrines). In the case of a new elevation, metastases should be sought by new imaging. Monitoring must be carried out for at least ten years [42,43]. A genetic diagnosis should be offered to all patients. Some mutations are correlated with a high risk of malignancy and a poor prognosis, particularly those involving the SDHB gene [14]. The search for mutations on susceptibility genes specifies whether or not the paraganglioma is part of a hereditary framework. This should lead to extending the genetic investigation and screening first-degree relatives. In our patient, the paraganglioma is probably not part of a hereditary framework. Nevertheless, a diagnosis of hypertension in the father should lead to the performance of a dosage of urinary metanephrines and normetanephrines.

Conclusion

Paragangliomas are rare tumors, most often observed in young adults, long asymptomatic. Surgical excision is the only radical treatment. It is sometimes difficult and complex. It is made more effective thanks to recent advances in anesthesia and resuscitation by controlling hemodynamic variations induced by catecholamines and laparoscopic surgery. Histology is often non-contributory in determining benign or malignant character. Long-term clinical, biological and radiological monitoring is always necessary. The hereditary potential of PGl requires a systematic genetic investigation.

Conflicts of Interest

None of the authors have any conflicts of interest (financial or otherwise) to disclose.

Funding

The authors received no financial support for the research, authorship, and/or publication of this article.

Informed consent

The patient provided written informed consent for the publishing of this case report and any related pictures.

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