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Multiple Paragangliomas: A Diagnostic and Therapeutic Challenge

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Abstract **Case Report**

Multiple paragangliomas are rare neuroendocrine tumours, often associated with a family history, whose management remains complex. We report the case of a 30-year-old female patient presenting with recent arterial hypertension and a left lateral cervical mass. Imaging performed as part of the extension assessment revealed a carotid paraganglioma associated with several retroperitoneal lesions, confirmed by functional imaging and biological tests revealing catecholamine hypersecretion. Initial medical management was initiated prior to multidisciplinary therapeutic discussion. This case highlights the importance of a personalised approach integrating multimodal imaging, biological assessment and genetic analysis to optimise diagnosis, treatment and long-term follow-up.

Keywords: Multiple Paragangliomas, Hypertension, Catecholamines, Functional Imaging, Multidisciplinary Management, Genetics (SDHx), Prognosis, Long-Term Follow-Up.

INTRODUCTION

Multiple paragangliomas (MPGL) are rare and generally slow-growing tumours, the diagnosis and management of which can be complex. The most common locations are carotid and jugulo-tympanic, while aortic involvement remains exceptional. These multiple forms often occur in a familial context. Treatment is mainly based on surgical resection, but in multifocal forms, the therapeutic strategy must be individualised according to the location of the tumours, their progression and the patient's profile [1-4]. We report the case of a young patient followed up in the endocrinology department for MPGL in the neck and abdomen.

CASE REPORT

This is a 30-year-old female patient with a history of recently diagnosed hypertension who presented with a left lateral cervical swelling that had been developing for a month, associated with frontal headaches, tinnitus and palpitations, without Menard's triad. There were no signs suggestive of multiple endocrine neoplasia. Clinical examination revealed a blood pressure of 137/95 mmHg, normal heart rate, normal breathing, and a left lateral cervical mass, with no other notable abnormalities.

The cervical MRI revealed a hypervascularised left cervical mass measuring 28x26x43 mm, projecting from the left carotid bifurcation and causing significant mass effect on the common carotid artery, the bulb and both internal and external carotid arteries, which remain patent, suggesting a grade III carotid paraganglioma (PPGL) **(Figure 1)**.

As part of the assessment to locate other lesions, a Cervical–thoracic–abdominal–pelvic CT scan (CT C-TAP) was performed, revealing multiple masses: left cervical, retroperitoneal projecting over the right adrenal gland and in line with the lower pole of the right kidney, measuring 31 x 28 mm, 32 x 36 mm, 24 x 22 mm and 21 x 20 mm, respectively **(Figure 2)**.

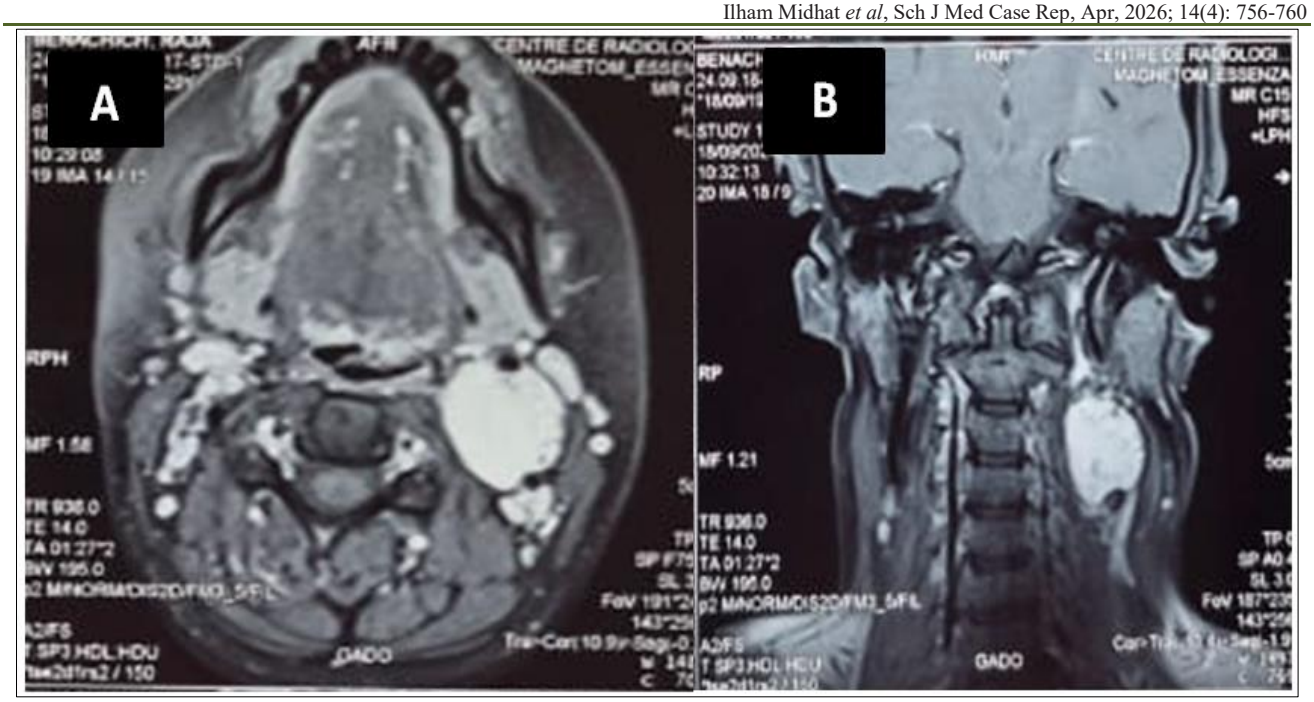

Figure 1: Axial (A) and coronal (B) sections of a cervical MRI revealing a well-defined left cervical mass measuring 28 x 26 x 43 mm, hypervascularised with projection of the left carotid bifurcation exerting a significant mass effect on the common carotid artery, the bulb and both internal and external carotid arteries, which remain patent, suggesting a grade III carotid paraganglioma

Figure 2 : Axial CT C-TAP images demonstrating, on abdominal sections, multiple retroperitoneal lesions projecting over the right adrenal gland and aligned with the lower pole of the right kidney, measuring 31 × 28 mm, 32 × 36 mm, 24 × 22 mm, and 21 × 20 mm, respectively

This was supplemented by somatostatin receptor scintigraphy (Octreoscan), which revealed an area of intense fixation in the left lateral cervical region, corresponding to a nodular mass measuring 32.4 x 27.9 mm, and visualization of multiple retroperitoneal nodular masses with well-defined contours moderately fixing the radiotracer, at the level of the right adrenal gland measuring 35.4 x 32.1 mm, and in the lower-inner part of the lower pole of the ipsilateral kidney (24 x 21.5 and 15.2 x 12.6 mm) **(Figure 3)**.

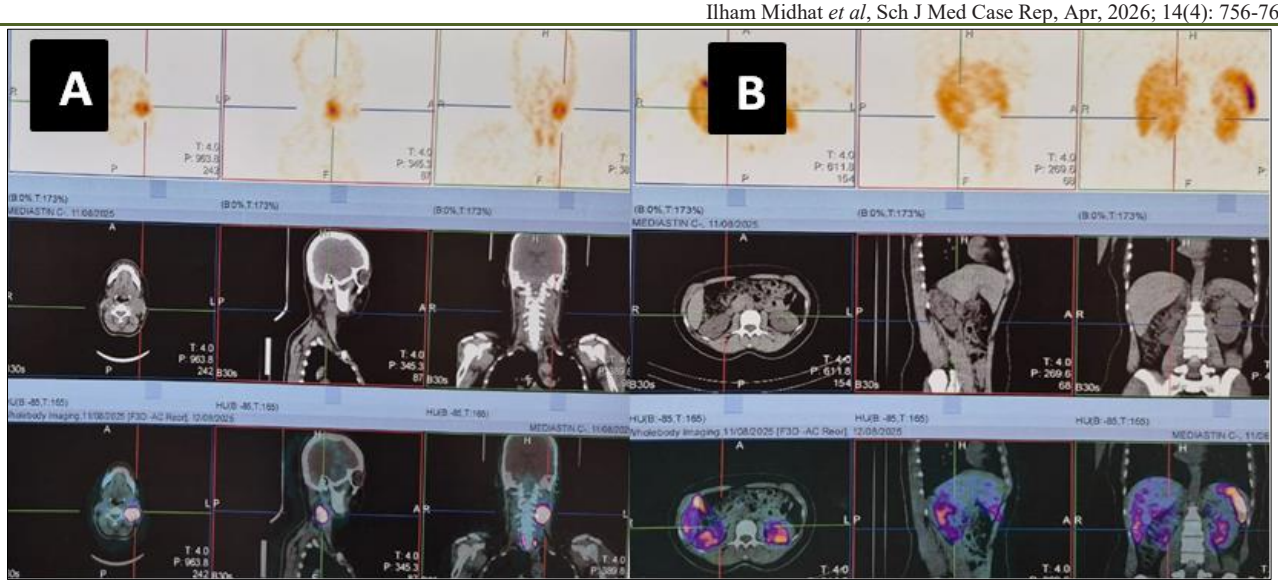

Figure 3: Somatostatin receptor scintigraphy (Octreoscan) combined with CT scan showing a left lateral cervical nodular mass with intense fixation, measuring 32.4 x 27.9 mm (A), Associated with multiple retroperitoneal nodular masses with well-defined contours, moderately fixing the radiotracer, at the level of the right adrenal gland measuring 35.4 x 32.1 mm and in the infero-internal part of the lower pole of the ipsilateral kidney (24 x 21.5 and 15.2 x 12.6 mm) (B)

Urinary methoxyl derivatives were elevated, with normetanephrines at 352 µg/24H (< 51) or 6.9xN, and 3-orthomethyldopamine at 308 µg/24h (<55) or 5.6 x N, and normal metanephrine levels. The blood pressure monitor revealed systolic-diastolic hypertension with systolic BP peaks reaching 172 mmHg. The assessment of the impact of hypertension was normal and the NEM assessment showed no abnormalities. Genetic testing for SDHx mutations or other genetic mutations was not performed, as it was not available in our context.

The patient was placed on alpha-blockers, and after discussion in a multidisciplinary consultation meeting, it was decided to perform excision of the intra-abdominal lesions and then discuss possible embolisation of the left carotid PPGL at a later date.

DISCUSSION

PPGL are rare neuroendocrine tumours, generally benign but with the potential of being malignant. They develop in the paraganglia, which are neuroectodermal structures derived from the neural crest, and can occur sporadically or hereditarily. MPGL is mainly observed in genetic forms, with a reported prevalence of up to 30-50% depending on the series. It most often occurs in a familial context, particularly in connection with mutations in the SDHD, SDHB, SDHC and SDHAF2 genes, and is characterised by bilateral and multiple tumour locations. These multiple lesions may be synchronous or metachronous [4].

The clinical presentation of pheochromocytomas (PCCs) and PPGL varies and depends on the tumour location, secretory nature, extent, malignant potential and hereditary or sporadic aetiology [5]. PPGLs originating from sympathetic paraganglia are most often secreting, causing typical adrenergic manifestations including headaches, sweating, palpitations, and persistent or paroxysmal arterial hypertension, with a high risk of cardiovascular complications in the absence of treatment [6-12].

Conversely, parasympathetic PPGL of the head and neck are generally non-functional, often asymptomatic, and frequently discovered incidentally or at an advanced stage due to a compressive effect [13-15].

The diagnosis of PCCs and PPGL involves clinical, biochemical, and radiological assessments. Plasma-free and urinary fractionated metanephrines are widely accepted for screening and follow-up, with plasma tests showing higher sensitivity [14, 5]. False positives can occur due to factors like diet, medications, stress, or lab errors, while false negatives may arise from small, asymptomatic tumors or biochemically silent types [13, 14]. Plasma 3-methoxytyramine (3MT) and chromogranin A measurements are recommended for specific tumor types, particularly those that are associated with mutations in the SDHB gene [13, 5]. Radiological imaging, such as CT and MRI, is crucial for tumor localization and staging. CT is preferred for initial localization due to its availability and spatial resolution, while MRI is better for extra-adrenal PGLs, pediatric patients, pregnant women, and long-term follow-up. Functional imaging, including techniques like 123/131I-MIBG SPECT/CT, 68Ga-DOTA-SSA PET/CT, and 18F-DOPA PET/CT, is used for inconclusive cases or suspected metastatic disease [14-19]. These imaging methods rely on molecular targets expressed on PPGL cells, such as norepinephrine transporters and somatostatin receptors, to detect tumors and guide treatment planning [5-18].

The management of PPGLs requires a multidisciplinary approach within an experienced medical team to define an optimal, individualized treatment strategy, taking into account both patient-related factors (such as age and overall health status) and tumor characteristics, including location, size, extent, biochemical profile, functional imaging findings, and, when appropriate, genetic background, while carefully weighing the benefits and risks of each therapeutic option [21, 5].

Surgery remains the cornerstone of treatment for the majority of localized PPGLs whenever feasible. In advanced or metastatic disease, management options include symptomatic therapy for functional tumors, such as alpha and beta-adrenergic blockade and catecholamine synthesis inhibitors, to prevent life-threatening complications, as well as systemic treatments (chemotherapy, targeted therapies, and somatostatin analogs), radiometabolic therapies, and selected locoregional interventions, including cytoreductive surgery, external beam radiotherapy, arterial embolization, cryotherapy, and radiofrequency ablation [13-16].

Due to the high prevalence of SDHx mutations in MPGL, genetic screening is recommended in patients with multiple locations, early age at diagnosis, or extra-adrenal involvement. This approach allows for more accurate prognostic stratification, adaptation of monitoring procedures, and organisation of appropriate family genetic counselling [6]. Furthermore, although malignant forms of PPGL are rare, they are mainly reported in the context of SDHB mutations.

Other genetic alterations, which are more rarely involved, may also be associated with multiple forms, in particular mutations in the VHL (von Hippel-Lindau disease), RET (multiple endocrine neoplasia type 2, most often responsible for bilateral PCCs), NF1, TMEM127 and MAX [13-25].

Long-term follow-up of at least 10 years is recommended for all patients after resection of PPGLs, as the risk of tumor recurrence persists in all cases. In patients with hereditary disease or those at high risk of metastatic progression, surveillance should be intensified and extended to lifelong follow-up [21]. Although no standardized follow-up protocols are currently established, monitoring should include periodic biochemical assessment and imaging studies. Whenever feasible, follow-up should be conducted within a multidisciplinary setting at a tertiary referral center [26].

Conclusion

MPGL are rare neuroendocrine tumours, most often benign and slow-growing, predominantly located in the carotid region. Clinical presentation varies depending on location, size and secretory nature. Diagnosis is based on morphological and functional imaging to assess tumour extent, with measurement of urinary methoxy derivatives to determine secretory character. Surgery is the standard treatment, with an individualised strategy, sometimes in several staged procedures, to limit postoperative morbidity. Other therapeutic options may be proposed depending on the location, operability and malignant or recurrent nature of the lesion. Genetic evaluation and prolonged monitoring are essential due to the risk of multifocality and malignant progression.

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