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Retroperitoneal Functional Paraganglioma: A Case Report and Review of the Literature

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ABSTRACT

Retroperitoneal paragangliomas are rare neuroendocrine tumors. They are defined as extra-adrenal chromaffin tumors and account for around 1/5th of all chromaffin tumors. They are often asymptomatic and can reach significant dimensions. We report the case of a 52-year-old female patient who presented with abdominal pain. Thoraco-abdomino-pelvic computed tomography (TAP CT) showed a right retroperitoneal tumor, urinary methoxylates were elevated. After adequate pharmacological preparation, the patient underwent surgery without complications. Blood pressure and urinary catecholamines were normal after surgery.

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KEYWORDS:

Paraganglioma, surgery, treatment, retroperitoneal functional paraganglioma.

INTRODUCTION

Paragangliomas are neuroendocrine tumors of neuroectodermal origin that can develop at the expense of sympathetic ganglia located along the spinal column in the thorax, abdomen, pelvis and genitourinary tract; they also occur in the cervical region and in parasympathetic ganglia located mainly in the cervical region and skull base (carotid, vagal, tympanic or jugular glomus)[1]. Retroperitoneal paragangliomas are very rare tumors, less frequent than other localizations (head, neck) [2]. They are often asymptomatic and can grow to considerable size. They require multidisciplinary management, but only surgical treatment is curative [3].

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CAS REPORT

Patient, 52 years old, with no particular pathological history, she reported pain in the right hypochondrium radiating to the hemicycle, associated with hot flushes without Menard's triad. Clinical examination revealed a conscious patient with normocolored conjunctiva, hypertensive at 149/94 mmhg, normocardiac at 79 bpm, eupneic at 18 cpm, overweight with pathological waist circumference, a homogeneous goiter without palpable nodule, pigmented lentiginous spots with multiple naevi on the abdomen, back and inguinal region.

Biological workup showed elevated 24h urinary methoxylates with normetanephrine at 80.98 µmol/24h (0.4 - 2.1), and metanephrine at 6.34 µmol/24h (0.2- 1) against normal 24h creatinuria.TAP CT revealed a right upper retroperitoneal mass separated from the right adrenal gland by the inferior vena cava, measuring 82×70×47 mm in tissue density (43UH) and moderately enhanced after contrast injection.

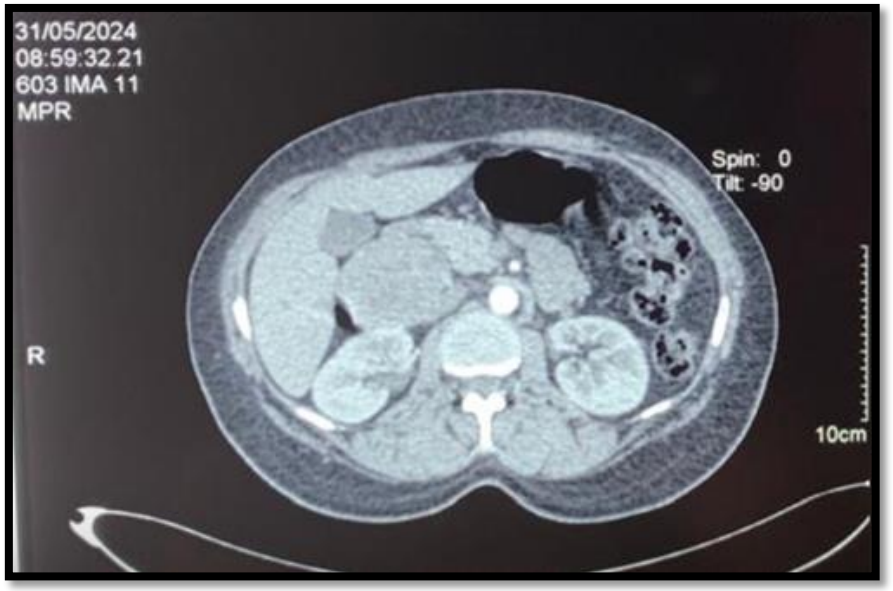

Figure 1: CT section showing the retroperitoneal mass above the right renal pedicle, measuring 82×70×47 mm.

After surgical preparation with 1 mg alpha blocker and rehydration, the patient underwent laparoscopic surgery, with a simple postoperative course. Histological examination revealed a tumour proliferation with a cord-like pattern and neuroendocrine architecture. Morphological analysis showed a moderately differentiated paraganglioma with a Gapp score of 6.

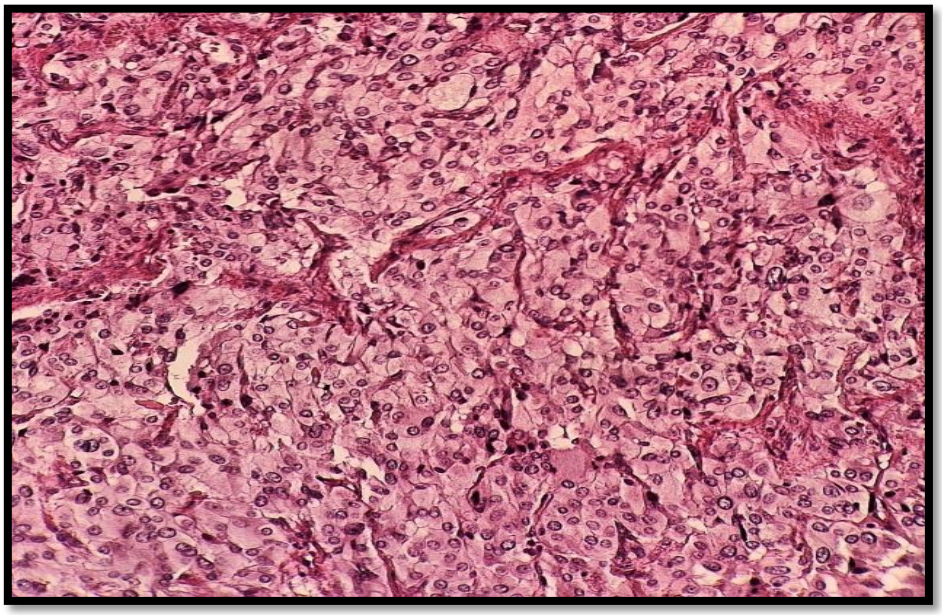

Figure 2: Showing endocrinoid tumor proliferation, the tumor cells are medium to large in size, with anisokaryotic nuclei with vesicular chromatin, and abundant, finely granular cytoplasm.

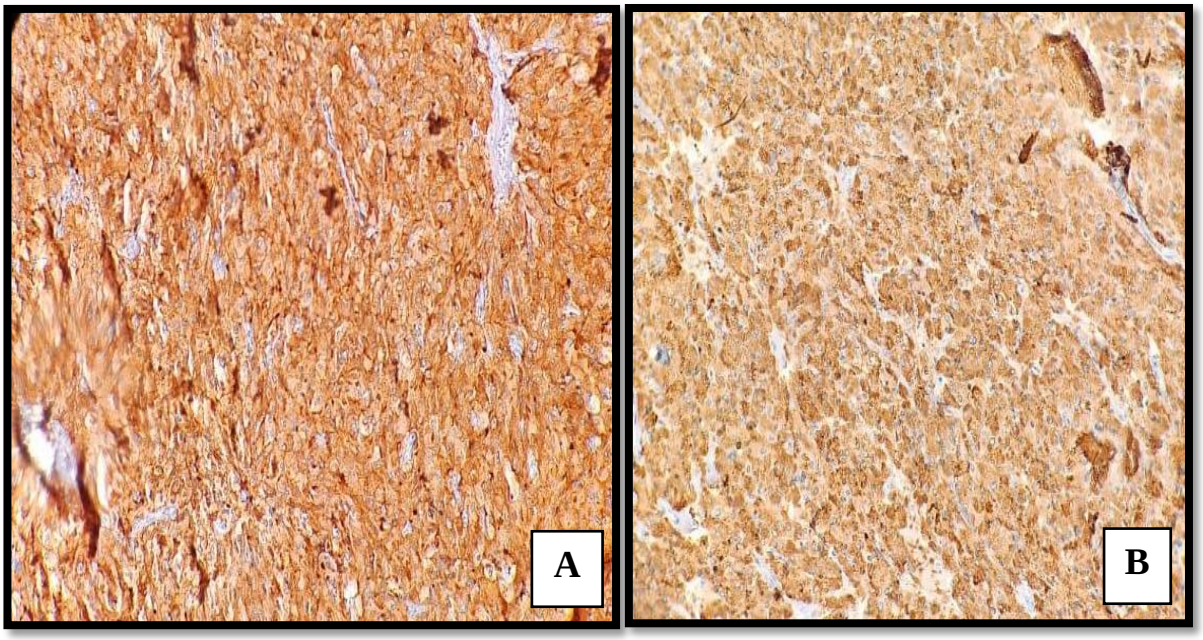

Figure 3: A Moderate and diffuse granular cytoplasmic expression of anti-Synaptophysin Ac B Moderate and diffuse granular cytoplasmic expression of anti-chromgranin Ac

Post-operative follow-up showed correct blood pressure and normal urinary methoxylates.

DISCUSSION

Paragangliomas represent a rare entity, accounting for around 1/5th of chromaffin tumors, and are secretory in 60% of cases. They occur at an earlier age, from 10 to 30 years, but are more frequent in young adults [4]. They are generally characterized by symptoms associated with hypersecretion of catecholamines and their metabolites. Paraganglioma patients generally present with persistent or paroxysmal hypertension, sweating, palpitations, headaches, anxiety, etc. The sudden release of catecholamines can be life-threatening, causing pulmonary oedema, alveolar haemorrhage and cardiovascular events such as cerebral haemorrhage, hypertensive crises, cardiac arrhythmias and myocardial ischemia [5 ,7]. Almost 40% of paragangliomas are non-secretory, which explains the absence of the above-mentioned symptoms and makes diagnosis difficult [6].

Positive preoperative diagnosis includes assessment of endocrine secretions by catecholamine assay, CT scan or magnetic resonance imaging (MRI), which is necessary for rapid localization of PGLs. CT is less sensitive than MRI [8]. Functional imaging is widely applied to assess local extension or multifocality, and to exclude metastases. Metaiodobenzylguanidine (MIBG) scintigraphy is a sensitive state-of-the-art technique, particularly when CT and MRI findings are negative or equivocal, and is particularly useful for diagnosing extra-adrenal tumors and metastases [12]. Genetic studies are strongly recommended for paraganglioma patients, in order to discover the genetic mutations that cause the disease. Revolutionary advances in genetics have already led to improved understanding and new discoveries with the classification of PPGLs into specific molecular groups [13][14]. Our patient was 52 years old and had a negative family history. We also strongly advised the patient to undergo genetic testing.

Preoperative pharmacological preparation with alpha-adrenergics for 2 to 4 weeks to prevent possible intraoperative catecholamine release, which can lead to hypertensive crisis, cardiac arrhythmia, pulmonary edema and cardiac ischemia due to tumor manipulation; a hypotensive crisis can even occur after tumor removal [9]. Radical surgery is the mainstay of treatment, with radical resection in 75% of cases. The choice of surgical procedure between conventional and laparoscopic approach is mainly related to the size of the tumour [10].

Complementary therapies, such as chemotherapy with a combination of Decarbazine, Vincristine and Cyclophosphamide, and external radiotherapy could find their place in metastatic forms, with a positive response in around 50% of cases, but without significantly influencing prognosis. Only surgical excision offers significant improvement, with a recurrence-free survival rate of 75% at 5 years and 45% at 10 years [11].

CONCLUSION

Retroperitoneal paragangliomas are neuroendocrine tumors developed at the expense of the parasympathetic nervous system. They are often asymptomatic and can reach large dimensions. Their management must be multidisciplinary, but only surgical treatment is curative. Complementary therapies, such as chemotherapy or external radiotherapy, may have a role to play in metastatic forms, but without significantly influencing prognosis.

DECLARATION OF PATIENT CONSENT

The authors certify that they have obtained the appropriate patient consent form. In this form, the patient gave her consent for her clinical and paraclinical information to be reported in the journal.

AUTHOR CONTRIBUTIONS

All authors contributed to the writing of this article.

CONFLICTS OF INTEREST

There are no conflicts of interest.

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