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Intraoperative Corrected Diagnosis of a Right Renal Para-Hilar Paraganglioma, Taken Preoperatively for a Renal Tumor

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Abstract **Case Report**

Nonfunctional retroperitoneal paragangliomas are rare tumors. They are defined as extra-adrenal chromaffin tumors and represent about 20% of chromaffin tumors. They are often asymptomatic and can grow to large dimensions. We report the observation of a 67-year-old patient admitted to the operating room for right total nephrectomy for right hilar renal tumor and whose intraoperative dissection of the renal pedicle could demonstrate an exorenal mass. Anatomopathological analysis was in favor of a paraganglioma. Malignant forms, more frequent than benign forms, present a locoregional invasion and metastasize late. The management of paragangliomas must be multidisciplinary but only surgical treatment is curative. However, there is no consensus on the usefulness of complementary therapies, which can nevertheless be used as a symptomatic adjunct. Paragangliomas are genetic in 25% of cases. A genetic investigation must be systematically proposed.

Keywords: Paraganglioma, nephrectomy, redirection of surgical indication.

BACKGROUND

Our case is interesting because the patient was initially admitted for a right nephrectomy. It was intraoperatively that the surgical dissection was able to clear the exorenal mass and that the surgical technique had changed.

Case Presentation

INTRODUCTION

Paragangliomas or extra-adrenal pheochromocytomas are rare (1/30,000) neuroendocrine tumors that develop from chromaffin cells derived from embryonic neural crests. They can be located between the base of the skull and the floor of the pelvis. They develop at the expense of the parasympathetic (head and neck paraganglioma) or sympathetic (thoracic, abdominal or pelvic paraganglioma) autonomic nervous system [1]. Non-functional retroperitoneal paragangliomas are rare tumors [2, 3] and are less frequent than other localizations (head, neck) [2]. They are often asymptomatic and can reach significant dimensions. The management of paragangliomas must be multidisciplinary but only surgical treatment is curative.

Observation

Mr B.A. was 67 years old and had a history of type 2 diabetes with insulin and benign prostatic hypertrophy with alpha-blocker and phytotherapy. Accusing a worsening of the signs of the lower urinary tract under treatment, then an abdomino-pelvic ultrasound requested came back in favor of a right posterior medio-renal tissue mass. A complementary CT scan was then requested, and came back in favor of a right renal hilar tumor mass with exorenal development whose appearance was suggestive of a clear cell carcinoma (Figure 1). The patient was staffed and the decision of the staff was to perform a complementary right renal magnetic resonance imaging. The latter showed a clearly malignant lesion process of the right kidney (Figure 2).

A preanesthetic consultation was requested and the patient was admitted to the operating room for a right renal nephrectomy. An open right subcostal surgery was performed to properly control the renal pedicle. Upon careful dissection of the pedicle, the mass was noted to be well exposed and in intimate contact with the right renal vein. After a quick discussion between surgeons, it was suggested that it may well be an extra-renal mass. The theory of a secretory

paraganglioma was plausible, for which the feasibility of the procedure was discussed with the anesthesiologist on the ward. It was decided to dissect the mass well despite the difficulty of the operating time. The dissection was successful and the mass was removed intact and without any anesthetic complications.

The anatomopathological analysis came back in favor of a paraganglioma of rather low aggressiveness, PASS score < 3 (Figure 3, 4)

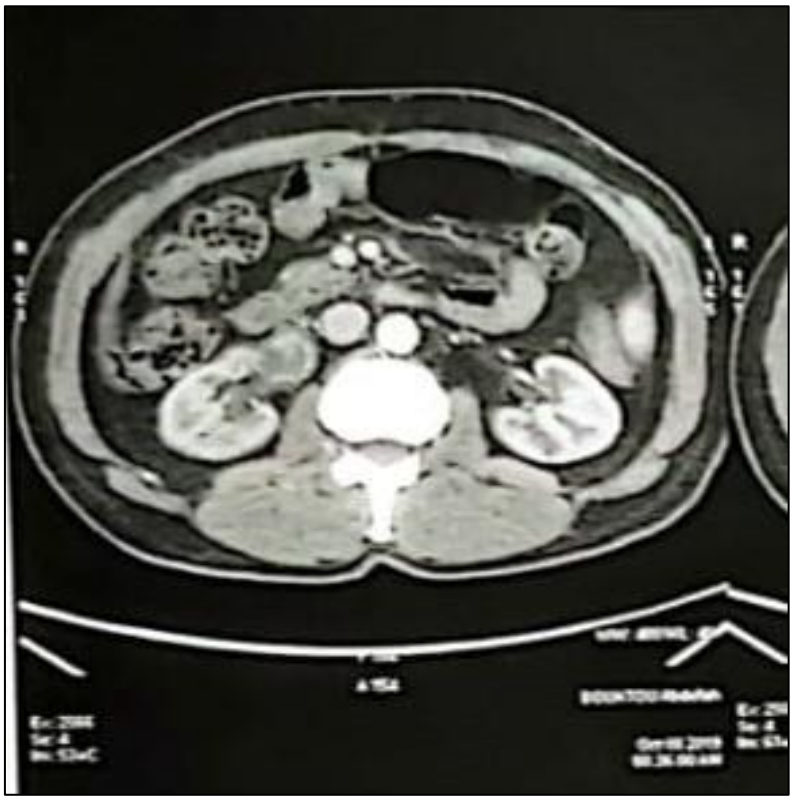

Figure 1: Right renal hilar tumor mass with exorenal development suggestive of clear cell carcinoma

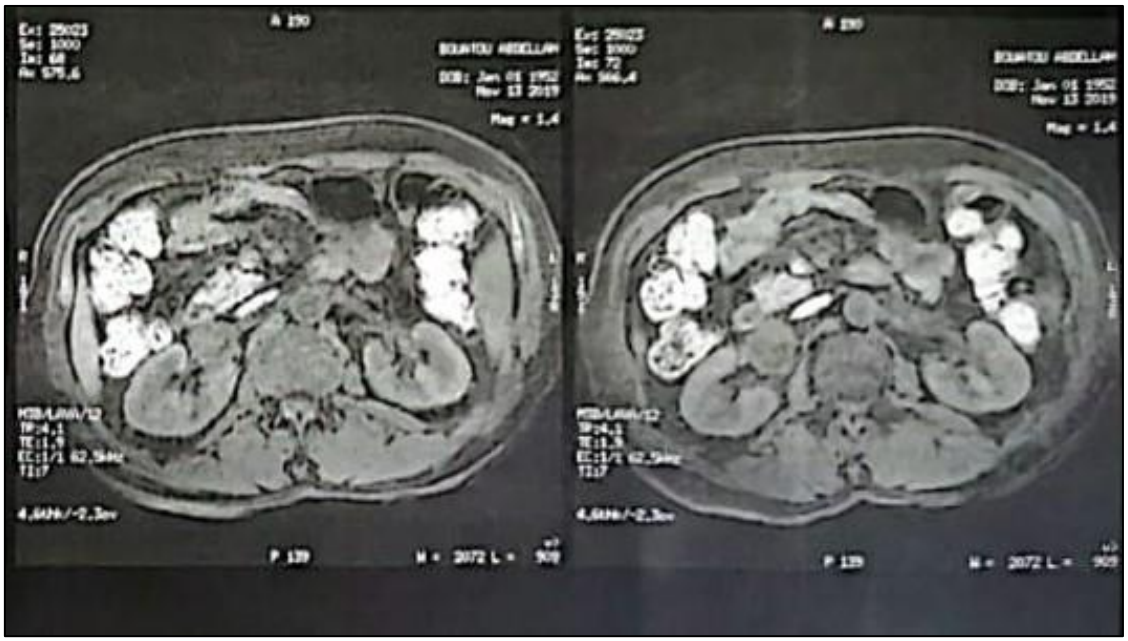

Figure 2: Lesional process of the right renal hilum measuring 31 x 27 x 50 mm, with heterogeneous signal, enhanced after gadolinium injection

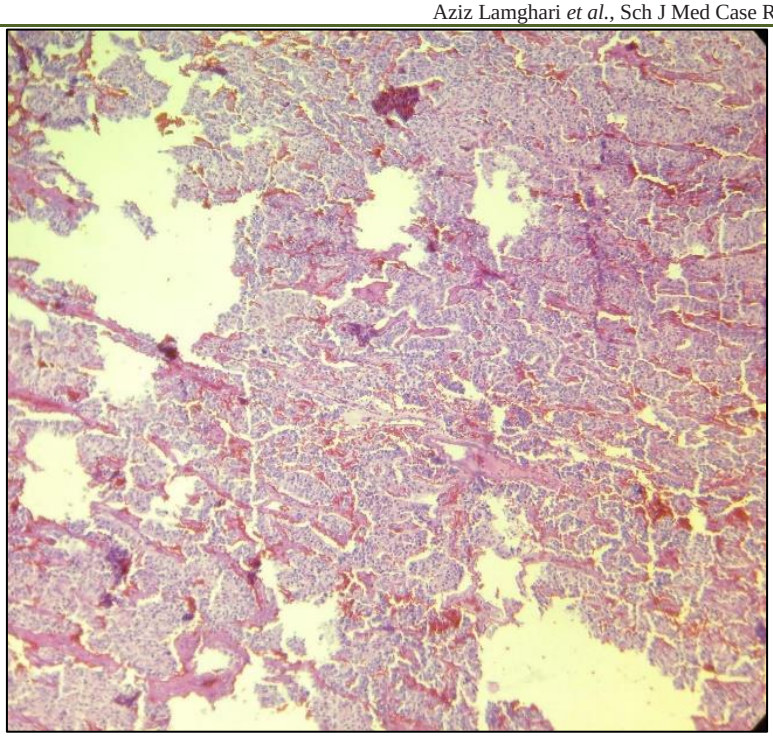

Figure 3: Tumor proliferation of neuroendocrine architecture made of cords separated by a fibro-vascular stroma (HEx100)

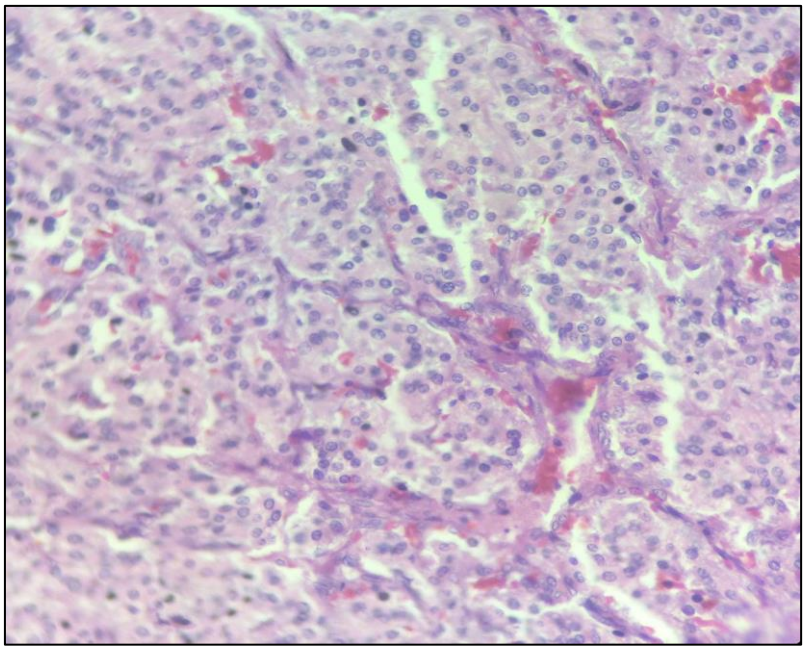

Figure 4: Cells have eosinophilic granular cytoplasm and nuclei with salt and pepper chromatin (HEx400)

DISCUSSION

Primary retroperitoneal tumors (PRT) in adults are most often malignant. They represent less than 1% of tumors. They are often discovered late in life, even though they reach a very large size. Tumor involvement of the retroperitoneum is heterogeneous. It is composed of 80% of malignant lesions of which more than half are sarcomas. The prognosis of these RPTs depends on their histological grade and complete surgical resection. The risk of local recurrence is greater than the risk of metastasis.

PRTs are classified according to their embryonic origin, thus paragangliomas are ectodermal tumors of neurogenic origin and are defined as extra-adrenal chromaffin tumors. Extra-adrenal pheochromocytomas or paragangliomas represent about 20% of chromaffin tumors and are a rare entity and are secretory in 60% of cases [4].

Embryologically, chromaffin cells extend from the base of the skull to the epididymis. Most of them involute; only those in the adrenal gland and retroperitoneal and thoracic ganglion chains persist. Extra-adrenal pheochromocytomas or paragangliomas account for 18% of chromaffin tumors. They occur at an earlier age, 10 to 30 years, but are more frequent in young adults [4]. They are multifocal in 15 to 24% of cases. They are much more often malignant than intra-adrenal pheochromocytomas, about 40% versus 10%. Malignant forms occur earlier than benign forms and are characterized by local (lymphatic type) or distant (invasion of the lung, bone, liver) invasion in 30% of cases [3, 4]. They predominantly secrete noradrenaline, often exclusively. Their origin is genetic, which corresponds well to their multi-centricity and metachronous recurrences in other sites. Only 1% of paragangliomas of the head and neck secrete catecholamines, whereas those of thoracic, abdominal, retroperitoneal or pelvic location secrete more often [5]. Eleven cases of pulmonary paragangliomas have been published. Finally, only two cases of paragangliomas or pheochromocytomas primarily intrahepatic have been described. Pheochromocytoma does not appear to be only a tumor of the adrenal gland, but rather a disease of the chromaffin system. The genetic origin is evoked more when the tumors are multiple, while the malignancy is higher when the tumor is ectopic.

Retroperitoneal forms are most often isolated [3,4]. They are sometimes associated with other pathologies, notably the Carney triad [6], with multiple endocrine neoplasia type 2 [3] and neurofibromatosis type 1. Non-functional retroperitoneal paragangliomas are characterized by their asymptomatic appearance (absence of arterial hypertension) and normal blood and urine catecholamine levels [3] and a clinical latency most often [4], sometimes non-specific signs are found [4].

The positive preoperative diagnosis is biological [7]. Abdominal ultrasonography shows an oval, solid, well-limited mass with numerous central cystic-like formations [3,6]. Abdominal CT shows the characteristics of the tumor, in particular its retroperitoneal location, its size, its single or multiple nature, with a search for locoregional and distant invasion. The most frequent appearance is that of a round or oval solid mass, homogeneous, but which may be cystic or necrotic in its center or calcified [3,8,9], whereas MRI is the examination of choice for the diagnosis and morphological assessment of the lesions. Meta-iodobenzylguanidine scintigraphy is of little interest preoperatively, but has a major role in monitoring.

The diagnosis of certainty is histological [3,4], with the appearance of a huge rounded tumor, encapsulated with a firm, elastic, highly vascularized consistency, but it is immunohistochemistry that makes it possible to affirm the diagnosis; however, there are no histological criteria that can distinguish between the benignity and malignancy of the tumor [8]. A good management of paraganglioma requires a precise morphological assessment, given the vascular complexity of these tumors, hence the interest in using angioscan. Radical surgery is the mainstay of treatment with radical resection in 75% of cases [4].

The choice of surgical procedure between the conventional and laparoscopic approach remains very controversial in view of the undeniable adverse effects of laparoscopy. Complementary therapies, such as chemotherapy or external radiotherapy, could find their place in metastatic forms with a positive response in about 50% of cases, but without significantly influencing the prognosis. Surgical excision is the only means of significant improvement, with a recurrence-free survival rate of 75% at 5 years and 45% at 10 years [4]. The average survival is about 3 years in metastatic forms and 4 years in case of incomplete excision [4].

CONCLUSION

Paragangliomas or extra-adrenal pheochromocytomas are neuroendocrine tumors that develop in the parasympathetic nervous system. Non-functional retroperitoneal paragangliomas are rare tumors. They are often asymptomatic and can reach significant dimensions. The management of paragangliomas must be multidisciplinary but only surgical treatment is curative. Complementary therapies, such as chemotherapy, external radiotherapy could find their place in metastatic forms but without influencing the prognosis significantly.

List of Abbreviations

PRT: Primary retroperitoneal tumors
Abdominal CT: Abdominal computed tomography
MRI: Magnetic Resonance Imaging

DECLARATIONS

Ethics approval and Consent to participate: Not applicable

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Authors' contributions:

AL, AE and MT analyzed and interpreted the patient data regarding the subject. MA and AA were a

major contributor in writing the manuscript. All authors read and approved the final manuscript.

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