Retroperitoneal Non-functioning Paraganglioma: A Case Report with Review of Literature

ABSTRACT

Background: Paragangliomas (PGs) are uncommon neoplasms that arise from primitive neural crest cells. These tumors are seldom detected and are typically difficult to identify and treat. They can either be functional or non-functional when they release catecholamines. A preoperative diagnosis of PGs is uncommon unless the tumor is functioning. A radiological diagnosis is made using computed tomography (CT) and magnetic resonance imaging (MRI). We describe the case of a male patient, age 41, who complained of abdominal pain and was later found to have a retroperitoneal tumor in the left anterior pararenal region. The tumor was entirely removed. Histological evaluation and immunohistochemical testing were used to confirm the diagnosis of PGs.

Case report: The purpose of this case is to describe an incidental finding of left retroperitoneal PGs in a young man who presented with left-sided abdominal pain. Additionally, we want to emphasize how crucial early diagnosis is and how dangerous these tumors can be.

Conclusion: Though they can potentially become locally invasive and metastasize, retroperitoneal PGs usually have a benign prognosis. Due to the potential for malignant transformation, surgical removal is the primary method of therapy for PGs.

Keywords: retroperitoneal tumors, paragangliomas, neuroendocrine tumors, immunohistochemistry.

1. INTRODUCTION

“PGs are relatively rare endocrine tumors that arise from paraganglionic tissue, a widely dispersed collection of specialized neural crest cells” [1]. “PGs, also known as extra-adrenal pheochromocytomas, account for 5–10% of pheochromocytomas and can occur in any position between the neck and the pelvis base” [2]. “The retroperitoneal PGs account for over 50% of all paragangliomas” [3]. “The most common site of extra-adrenal occurrence is intra-abdominal, usually within the para-aortic and perinephric spaces” [4]. It can occur at any age, most commonly in young adults, and tends to be sporadic.

However, they might be related to syndromes in approximately 30% of cases. They may be functional and result in symptoms of excess catecholamine production. Approximately 10–15% of such tumors are nonfunctional; in another 10%, hormone activity does not manifest clinically. They are frequently locally invasive and have a high incidence of local recurrence.

Various imaging techniques are essential to detecting these tumors.

The typical CT appearance of PGs is a well-limited, isodense mass with intense contrast enhancement. On MRI, the mass presents an iso- or hypo-signal in T1 and a hyper-signal in T2, with an early and intense enhancement. Thus, these tumors might be misconstrued as additional primary epithelial or mesenchymal abdominal malignancies. Since benign and malignant retroperitoneal PGs have the same histological appearance, the best predictor of malignancy is metastasis or recurrence.

The possibility of malignant transformation of PGs makes surgical excision the treatment of choice.

The present case describes the clinical presentation, radiological features, and histological features of nonfunctioning retroperitoneal PGs discovered during the investigation of abdominal pain.

2. CASE REPORT:

A 41-year-old male was referred with vague abdominal pain of 4 months duration. Further inquiry showed no history of hypertension, chest tightness, palpitations, anxiety, or weakness. He had undergone a ventral hernia repair a few years ago. He was taking no regular medications. No endocrine problems run in the family. His vitals remained stable during the hospital stay.

The physical examination revealed a palpable and tender mass in the upper left abdominal quadrant; the pain was sudden in onset and progressive in nature. The rest of the physical examination was otherwise normal.

Initial investigations, including biological assessments and X-rays, were all normal. However, abdominal CT was performed and revealed a well-defined retroperitoneal mass lateralized on the left, round-shaped, with regular contours, spontaneously hypodense and heterogeneous, intensely and heterogeneously enhanced after injection of contrast in the arterial phase, with a close connection to the anterior pararenal space and the psoas muscle, measuring 82*71*100 mm, without locoregional invasion, suggestive of liposarcoma or neuroendocrine tumors (Figure 1). On MRI, the mass presents an iso signal, or hypo signal, in T1 and a hyper signal in T2, with an early and intense enhancement. A diagnosis of GIST was made (Figure 2). These tumors might therefore be confused with other primary mesenchymal or epithelial abdominal tumors.

Through a midline laparotomy incision, after mobilization of the left colon, we found a vascularized retroperitoneal tumor of about 10 cm that was encapsulated and soft, with proximity to the left ureter and the psoas muscle. The lesion was wholly resected after a laborious ureterolysis (Figure 3). The lateral boundary of the dissection was at the left ureter (Figure 4).

There were no intraoperative or postoperative complications, and he was released from the hospital on the sixth postoperative day.

On macroscopic examination, the mass was encapsulated with a lobulated and central area of hemorrhage. Histological examination of the surgical specimen coupled with the immunohistochemical study was consistent with the diagnosis of PGs.

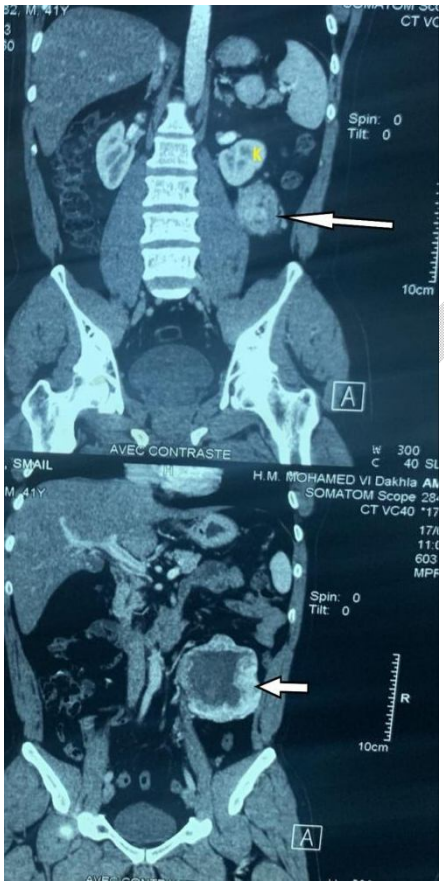

Figure 1. Abdominal CT scan with injection of intravenous contrast : well-defined retroperitoneal mass (white arrow)

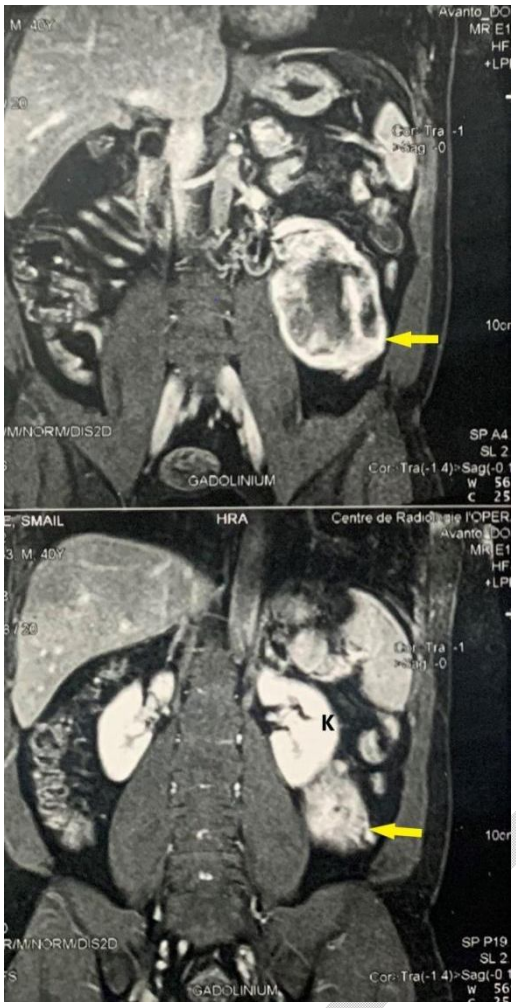

Figure 2. MRI image show the PGs (yellow arrow), with a close connection to the anterior pararenal space (K : the kidney)

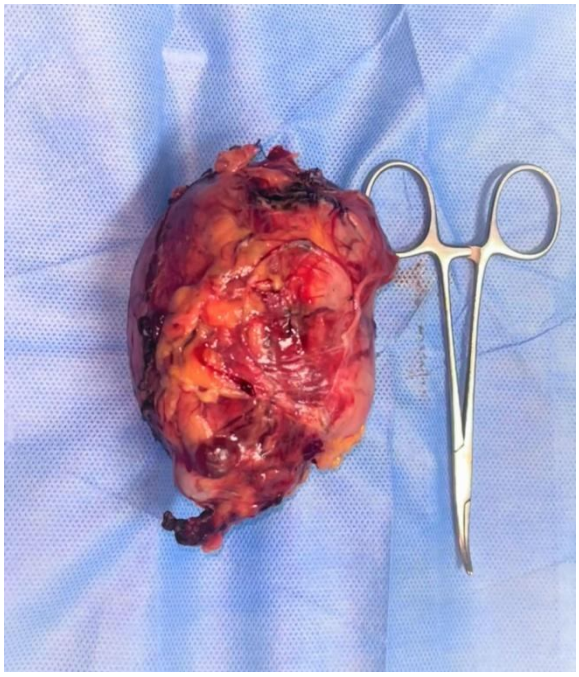

Figure 3. The excised PGs

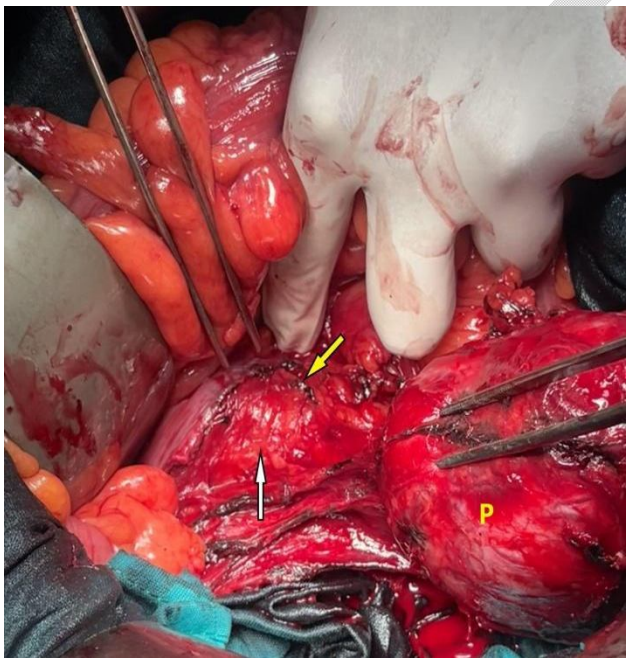

Figure 4. Operative photo : note the PGs (P) and close proximity of the ureter (yellow arrow) and the psoas muscle (white arrow)

3. DISCUSSION

“PGs are benign tumors of the sympathetic and parasympathetic paraganglia that arise from neural crest cells. Phaeochromocytomas, which arise from the adrenal medulla, account for nearly 80% of these tumors. Extra-adrenal paragangliomas (ten percent to twenty percent) develop from chromaffin tissue along the autonomic nerve system. As a result, they are found in the pelvis, head, neck, thorax, abdomen; and The abdomen is the most frequent location for sympathetic PGs, which are generally found in the organ of Zuckerkandl near the aortic bifurcation, which is consistent with the distribution of paraganglia initially described in the human embryo by Zuckerkandl in 1901” [6]. “These tumors are occasionally numerous or coupled with other PGs or tumors, such as gastrointestinal stromal tumors of the stomach and pulmonary chondromas, as a component of Carney's triad. The average patient age at diagnosis is around 40 years old, and men and women are equally affected. Many PGs are linked to disease-causing mutations or genetic disorders, such as VHL gene mutations” [7].
Patients with functioning PGs that create catecholamines may feel hypertension, flushing, tachycardia, palpitations, and anxiety [8], and they are easily diagnosed because urine catecholamines are increased. Those with nonfunctioning retroperitoneal PGs have an asymptomatic profile and normal catecholamine levels in the urine and blood. And as in our patient, it may be identified incidentally or manifest with compressive symptoms such as abdominal pain, or it may be linked with nausea, vomiting, abdominal distension, and weight loss. [9]

Unless the tumor is functioning, retroperitoneal PGs are seldom diagnosed. Because of their location, non-secreting nature, and sluggish development, they are clinically latent tumors. Although ultrasound can be utilized as a first-line inquiry, CT and MRI offer superior sensitivity. CT scans often reveal a heterogeneous, solid round, or oval mass with central necrosis or bleeding, calcification, and enhancement. Although imaging methods can be useful, the diagnosis of PGs can only be verified after rigorous histological and immunohistochemical investigation [10], as is the case in this instance. PGs are distinguished histologically by their extremely vascular appearance, with chief and sustentacular cells organized in zell-Ballen clusters. This abundant vascular tumor component explains the high contrast enhancement in CT or MRI imaging. Specific antibodies for neuroendocrine markers, such as synaptophysin, chromogranin, and S-100 protein, may also be utilized to confirm the diagnosis [11]. Due to the malignant potential of PGs, surgical excision is the only way to treat them. Resection is usually difficult since these highly vascular tumors are typically found near several important blood vessels. If a tumor is unresectable, efforts to shrink it via chemotherapy, radiation treatment, or embolization may be justified because surgery is the only way to cure it [12]. These supplementary treatments can provide a good response in 50% of instances while having no effect on the prognosis [13]. Surgery is the only way to dramatically improve prognosis, with a five- and ten-year survival rate of 75% and 45% without recurrence, respectively.

Extra-adrenal tumors are believed to be more likely to be malignant than adrenal tumors [14]. Metastasis can occur in the bone, liver, peritoneum, pelvis, cervical lymph nodes, ovaries, and lungs. Because PGs are uncommon tumors, substantial prospective trials are lacking, and all existing series have a low ability to identify significant differences in survival in patients with or without metastasis at initial presentation. In a study of 22 patients with extra-adrenal retroperitoneal PGs at Memorial Sloan-Kettering Cancer Center, the 5-year survival rate for tumors that were not removed was 19%, compared to 75% for tumors that were completely excised [15]. This is comparable with Cunningham et al.'s [16] 5-year survival rate of 73%. Metastasis may occur up to 7 years following resection. When metastasis develops, the average survival time is fewer than three years [15]. There was no difference in survival time between functional and nonfunctional paragangliomas. Postoperative CT, MRI, and PET scans are advised to identify malignant potential or disease progression. It should begin three months following surgery and be repeated every two years

for the first three years. For PGs, the recommended total time of follow-up is at least ten years, and if there is a significant risk of malignant PGs, the follow-up must be prolonged [17]. Despite their rarity, this case highlights the need to include extra-adrenal PGs in the differential diagnosis and therapy of retroperitoneal malignancies. Imaging investigations have greatly aided in the more accurate diagnosis of PGs. However, the gold standard of diagnosis remains pathological. Patients with retroperitoneal PGs must be followed for the rest of their lives since metastasis and recurrence are possible.

4. Conclusion

PGs of the retroperitoneum are an uncommon type of tumor with malignant potential that is difficult to diagnose and cure. Our case highlights the importance of including extra-adrenal PGs in the differential diagnosis of retroperitoneal tumors. The malignant types are distinguished by recurrence and metastasis. Catecholamine levels should be utilized to examine patients initially, followed by CT or MRI to pinpoint the main lesion. The ultimate diagnosis is based on immunohistochemistry results. For circumscribed malignancies, surgical excision remains the primary treatment. Advances in genetic testing and the identification of novel molecular markers have helped our knowledge of PGs. There is, however, no way to predict metastatic risk with certainty.

To identify early tumor recurrence, regular ultrasound and CT scans are required.

CONSENT

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.
Ethical Approval:

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

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