



Non-secreting Retroperitoneal Paraganglioma Mimicking a Para-Aortic Abscess: A Case Report and Review of the Literature

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Abstract

Non-secreting retroperitoneal paraganglioma is a rare entity that originates in chromaffin cells of the sympathetic and parasympathetic constituent of the autonomic nervous system. Its variable and nonspecific presentations and its infrequent incidence can lead to delayed diagnosis, hence the risk of its expansion into large sizes. Our paper describes a rare form of retroperitoneal paraganglioma mimicking a para-aortic abscess treated by laparoscopic surgery. We also refer to a literature review to discuss this entity's clinical, biological, radiological, and therapeutic features.

Keywords: Retroperitoneal paraganglioma; Pheochromocytoma; Mimicking abscess; Extra-adrenal pheochromocytoma; Blood cultures; Chromaffin cells; MIBG; Succinate dehydrogenase; Chromogranin

Introduction

The Paraganglioma (PGs) is a rare tumor that originates in chromaffin cells of the sympathetic and parasympathetic constituent of the autonomic nervous system. The retroperitoneal localization is the most frequent [1] and is for long asymptomatic, especially since 40% to 50% are non-functional. Herein, we illustrate a very unusual manifestation of a retroperitoneal PG mimicking a para-aortic abscess. It allowed us to make an early diagnosis of this insidious pathology. Our paper demonstrates that sepsis can be a rare but not an extraordinary inaugural clinical event.

Case Presentation

A 49-year-old female without a past medical history was admitted to the emergency department with fever and acute abdominal pain. The initial clinical evaluation revealed a body temperature of 38.6°C, a pulse rate of 78 beats per minute, a blood pressure of 110/60 mmHg, and a left flank rebound tenderness. Blood tests showed a hyperleukocytosis at 14.6/mcl (normal 4,500 to 11,000 leukocytes/microliter) and a high C-reactive protein 200.1 mg/dl (normal 0 to 0.8 mg/dl). Blood culture samples were taken before the administration of an empirical antibiotic consisting of amoxicillin/clavulanic acid. Three days later, they returned positive for *Escherichia coli*, sensitive to the initial antibiotic chosen, which were maintained. Computed Tomography (CT) scan showed a left para-aortic mass with a relatively low-attenuation central necrotic component and capsular ring enhancement in favor of an abscess (Figure 1). The patient had an excellent clinical outcome under antibiotics and was discharged from the hospital on the fourth day. A control contrast-enhanced CT scan was performed two months later and revealed a left para-aortic heterogenous mass evoking a retroperitoneal PG (Figure 2). MRI of the neck and chest CT scan were unremarkable. The plasmatic assay of metanephrine and normetanephrine were within normal limits. The patient underwent surgical laparoscopy with resection of the PG (Figure 3A) and had a satisfactory recovery before being discharged on the third day. The histopathological examination and immunohistochemical results yielded a PG, and tumor cells displayed immunoreactivity for neuroendocrine markers: Chromogranin and synaptophysin (Figure 3B). The follow-up at one year showed no recurrence of her condition.

Discussion

The PG or extra-adrenal pheochromocytoma is a rare catecholamine secreting tumor, which

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Figure 1: Left para-aortic mass capsular ring enhancement with contrast mimicking an abscess (orange arrow).

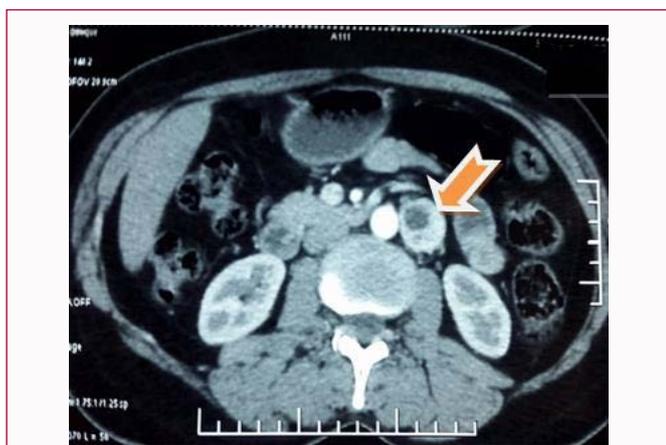


Figure 2: Para-aortic mass corresponding to a retroperitoneal paraganglioma (orange arrow).

arises in the extra-adrenal chromaffin cells. The retroperitoneal localization is the most frequent one and constitutes about 5% to 10% of all chromaffin cell tumors and originates in the para-aortic sympathetic system and urinary bladder, while only 5% are located in parasympathetic tissues in the thorax or head and neck [2]. A 10% to 30% of PGs are malignant [3]. This malignancy is defined by the presence of metastasis in a lymph node or other distant sites [4]. PG is a sporadic disease in 75% of cases. However, a recent paper suggested that more than 30% of pheochromocytomas have now been proved to be hereditary and associated with germline mutations [5], principally in the Succinate Dehydrogenase (SDH) gene (SDHA, SDHB, SDHC, SDHD, and SDHAF2). These tumors associated with SDHB mutation are more likely to arise in an intra-abdominal extra-adrenal location and have a very high risk of metastasis [6]. Genetic testing in hereditary PG is highly recommended since it allows the screening and detection of the disease in a pre-symptomatic phase, permitting an early diagnosis, management, and a follow-up of these patients to avoid missing a recurrence or metastasis [7]. The retroperitoneal PG condition remains asymptomatic for a long time until it reaches enormous sizes. Currently, and thanks to the development of imaging, more than half of PGs are accidentally diagnosed as incidentalomas. However, in the other cases, as described in Xiao-Ke Ji's et al. paper, the diagnosis is made after some frequent clinical signs as an

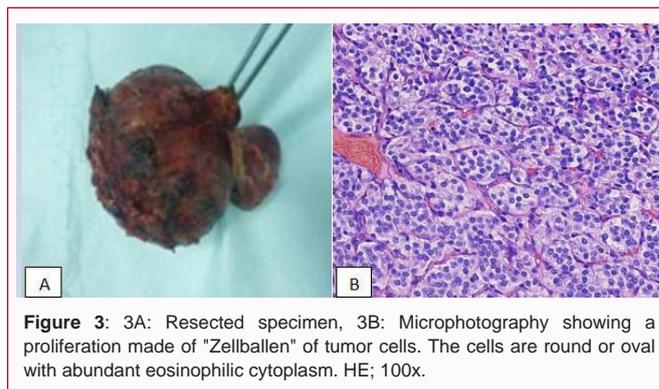


Figure 3: 3A: Resected specimen, 3B: Microphotography showing a proliferation made of "Zellballen" of tumor cells. The cells are round or oval with abundant eosinophilic cytoplasm. HE; 100x.

abdominal mass, hypertension, and abdominal pain [1]. However, none of his patients had sepsis or abscess as a revealing symptom. This rare inaugural clinical manifestation was only described twice in the literature (Table 1). The first case reported was in 2008 by Yau KK's paper [8]. An acute abdomen was mistaken for appendicitis but turned out to be a ruptured retroperitoneal PG. The second case was reported in 2010 by Arrabal-Polo MA [9], which described a spontaneous retroperitoneal abscess as a revealing PG's symptom. This is also our patient's case, where an early diagnosis was made thanks to this unusual symptomatology. Biochemical diagnosis of PG is based on either the dosage of plasmatic or urinary metanephrine and normetanephrine. No consensus is established until now, but the endocrine society recommends using plasmatic measure considering that the specificity and sensitivity are higher [10]. PG's initial diagnosis can be performed with thoracic, abdominal, and pelvic CT scan with a cervical Magnetic Resonance Imaging (MRI), and when the diagnosis is established, a 123-I Meta-Iodobenzyl-Guanidine (MIBG) scan is essential. If metastasis is suspected (presence of an SDH-B gene mutation), an FDG PET/CT is recommended by the European Society of Endocrinology Clinical Practice Guideline [4]. However, a recent paper showed that the Gallium-68 DOTATATE (or Ga-68 DOTATATE PET (positron emission tomography)) scans are superior for the detection of early metastatic disease to the other imaging modalities [5]. The curative treatment of retroperitoneal PG is surgery. The tumor's resection can be preceded either by laparoscopy or laparotomy after preoperative management to control hypertension and avoid cardiovascular events, using alpha and beta-adrenergic blockade, 7 to 15 days before the surgery. The Endocrine Society recommends using alpha-adrenergic blockade with prolonged action. The phenoxybenzamine is the standard molecule used with a progressive dose. If the treatment is insufficient, the addition of an alpha blockade and Calcium channel blocker can be used [2,11]. Thus far, there is no consensus for the management of metastatic PG. The treatment modalities available in our therapeutic arsenal are numerous, such as chemotherapy, MIBG therapy, external irradiation for bone metastases, and targeted molecular therapy like tyrosine kinase inhibitor [3]. The duration of follow-up recommended is at least ten years for PGs to screen the disease's recurrence and metastasis. Though, if there is a high risk of malignant PG like a mutation of SDH-B, the follow-up must be extended [4]. For the catecholamine-secreting PG, the follow-up consists of a plasmatic or urinary assay of metanephrine and normetanephrine 2 to 6 weeks after surgery, then once a year [4,10]. For the non-secreting tumors, the follow-up consists of imaging every 1 to 2 years or an assay of chromogranin yearly if it was positive initially [4,11].

Table 1: Comparison of the clinical, biological, radiological, and therapeutic features between the series published in the literature and our case.

Author (Year)	Miguel Angel Arrabal-Polo et al. [9]	Kwok-Kay Yau et al. [8]	Our case
Gender	Woman	Man	Woman
Age (years)	54	76	49
Medical history	No	Diabetes and hypertension	No
Abdominal pain	Yes	Yes	Yes
Temperature	37°C	-	38.6°C
Physical examination	Mass in the left hypochondrium	local peritonism in the right lower quadrant of the abdomen	left flank rebound tenderness
Leukocytosis	17.81 × 10 ⁹ /L	17.4 × 10 ⁹ /L	14.6 × 10 ⁹ /L
C-reactive protein mg/dl	19.65	-	200.1
Blood culture	Yes	No	Yes
Imaging	Retroperitoneal mass with a low attenuation compressed and displaced the left kidney, left suprarenal gland, and the tail of the pancreas	Hyper vascular tumor extending from the right renal hilum down to the level of the aortic bifurcation	Left para-aortic mass with a relatively low-attenuation central necrotic component and capsular ring enhancement
Plasmatic or urinary catecholamines	-	Normal	Normal
Treatment	Antibiotics and surgery	Surgery	Antibiotics and surgery
Histopathological examination	retroperitoneal PG	retroperitoneal PG	retroperitoneal PG
Positive immunohistochemical results	Vimentine, chromogranin A, synaptophysin, and PS100	-	Chromogranin Synaptophysin
Follow-up	-	CT scan at 6, 18, and 36 months showed no recurrence	No recurrence after 12 months

Conclusion

The non-secreting retroperitoneal PG is a rare entity often asymptomatic that could expand until getting large sizes, abscess or sepsis are rare but not exceptional revealing symptoms of this pathology.

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