Open Journal of Clinical & Medical Images

www.jclinmedimages.org

*Case Report*

*Open Access, Volume 3*

# Acute abdomen revealing bilateral retroperitoneal paragangliomas: A case report

***K Rifai[1,2]*; L Guissi[1]; N Moussaid[1]; M Babty[3]; T Karmouni[2,3]; H Iraqi[1,2]; MH Gharbi[1,2]***

*[1]Department of Endocrinology and Metabolic Diseases, Ibn Sina University Hospital of Rabat, Morocco.*

*[2]University of Medicine and Pharmacy, Mohammed V University, Rabat, Morocco.*

*[3]Department of Urology, Ibn Sina University Hospital of Rabat, Morocco.*

***Corresponding Author: Kaoutar Rifai**
University of Medicine and Pharmacy, Rabat, Morocco.
Email: kaoutar.rifai@um5r.ac.ma

Received: May 30, 2023

Accepted: Jul 06, 2023

Published: Jul 13, 2023

Archived: www.jclinmedimages.org



**Keywords:** Retroperitoneal paragangliomas; Hypertension; Acute abdomen.

## Abstract

**Background:** Paragangliomas are rare neuroendocrine tumors that develop from chromaffin cells derived from the embryonic neural crest.

**Results:** We report a unique case of a 23-year-old female patient who presented to the emergency department with an acute abdomen. The clinical examination revealed severe hypertension, diffuse abdominal tenderness without defensiveness or contracture. Laboratory results showed an elevated urinary normetanephrine level. Imaging revealed bilateral hypervascular retroperitoneal masses. The evolution was favorable after tumor resection with normalization of blood pressure. Anatomopathological study confirmed the diagnosis of paragangliomas.

**Conclusion:** Retroperitoneal paragangliomas can manifest as an acute abdomen. The diagnosis should be considered in the presence of any hypervascular retroperitoneal mass.

## Introduction

Paragangliomas are neuroendocrine tumours of neuroectodermal origin arising from the sympathetic and parasympathetic ganglia [1]. They usually present as a single unilateral tumor [2]. Retroperitoneal paragangliomas are rare tumours and less common than other localisations (head, neck) [3].

We report a rare case of a 23-year-old female patient with bilateral retroperitoneal paragangliomas initially manifesting as acute abdomen.

## Observation

A 23-year-old woman without a personal or family pathological history came to the emergency department with intense abdominal pain, nausea, and vomiting. On physical examination, blood pressure was elevated at 200/120 mmHg, body temperature was 36.5°C, and an electrocardiogram showed sinus tachycardia at 118 beats/minute. The patient reported the classic Menard's triad "headache-palpitations-sweating". Diffuse abdominal tenderness was noted but there was no defensiveness or contracture.

The diagnosis of pheochromocytoma was suspected because of the association of hypertensive peak and Menard's triad. The 24-hour urinary methoxylated derivatives returned high, urinary normetanephrine and metanephrine were raised to 69.3 µmol/24h (reference range: 0.40-2.10) and 1.26 µmol/24 h (reference range: 0.20-1.00), respectively. Abdominopelvic Computed Tomography (CT) revealed two hypervascular retroperitoneal masses in the left lateroaortic region (size: 80 x 60 x 45 mm) and in the right lateroaortic and precaval regions (size: 52 x 38 x 15 mm) (Figure 1). MIBG scintigraphy showed intense uptake by retroperitoneal masses suggesting paragangliomas, with no other distant suspect focus of hyperfixation (Figure 2).

Genetic investigation revealed no similar cases in the family and no associated hereditary conditions (von Hippel Lindau disease, neurofibromatosis type 1, and multiple endocrine neoplasia type 2). Our patient was tested negative for the succinate dehydrogenase B subunit (SDHB) mutation.

The patient underwent laparoscopic resection of paragangliomas (Figure 3) after 15 days of medical preparation with alpha-blocker (Doxazosin 4 mg/day with gradual titration) and beta-blocker (bisoprolol). The postoperative course was uneventful. Histopathologic examination of the surgical sample confirmed the diagnosis of bifocal paraganglioma measuring 8 × 5.5 × 3 cm and 5.8 × 2.5 × 2 cm with a total weight of 110 grammes. There was no evidence of vascular emboli or capsular invasion. The evolution was marked by the disappearance of Menard's triad and arterial hypertension. The 24-hour urinary methoxylated derivatives were normal two weeks after the operation.

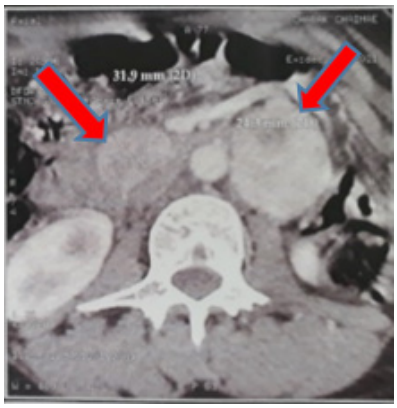

**Figure 1:** Retroperitoneal paragangliomas on CT scan.

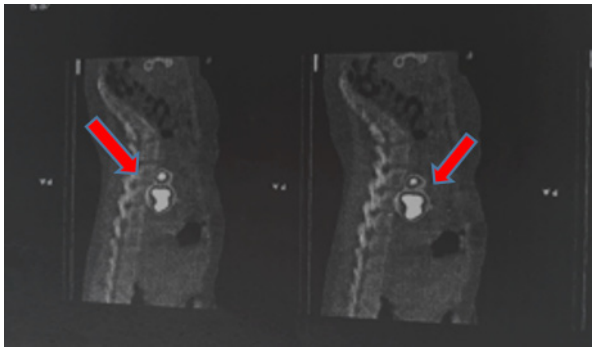

**Figure 2:** Retroperitoneal paragangliomas on MIBG scintigraphy.

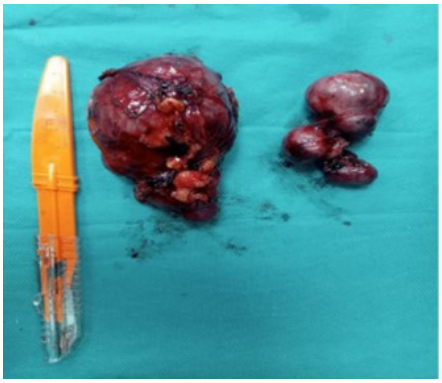

**Figure 3:** Surgical specimen of paragangliomas.

## Discussion

Our case illustrates a rare cause of secondary hypertension in young people, with the particularity of a bilateral retroperitoneal location of paragangliomas as well as the unusual mode of revelation by an acute abdomen.

It is a rare disease; the prevalence of paragangliomas in a population of hypertensive patients is estimated between 0.1 and 0.6% [1]. In fact, rare cases have been reported in the literature.

Paragangliomas (PGLs) can be located anywhere from the base of the skull to the pelvic region. They develop at the expense of the parasympathetic (head and neck paragangliomas) or sympathetic (thoracic, abdominal or pelvic paraganglioma) autonomic nervous system [4].

Thoracoabdominopelvic PGLs are functional in about 85% of cases [1]. They mainly secrete noradrenaline, often exclusively [3], as was the case for our patient. On the contrary, paragangliomas of the skull base and the head and neck region are non functional in 90% of cases [1]. These tumors can occur at any age, in both women and men.

The symptomatology of PPGLs may be related to catecholamine hypersecretion and/or to tumor mass effect. Arterial hypertension is the most frequent clinical sign, the classical triad of headaches, palpitations, and sweating is found in 60-90% of cases. Other symptoms are less suggestive: weight loss, anxiety, pallor, nausea. Very rare cases of shock with multi-visceral failure and hyperthermia have also been described [1]. Our patient presented with an acute abdomen as the initial manifestation of retroperitoneal paragangliomas. This clinical presentation is extremely rare and complex. However, retroperitoneal tumor accompanied by infection, hemorrhage or compression can lead to an acute abdomen [5].

The diagnosis of functional PGL is established by the *demonstration* of increased secretion of catecholamines in their methoxylated derivatives. They are measured on 24-hour urine or a plasma assay in case of renal failure. The diagnosis is highly probable if their concentration is greater than three times the upper limit of normal [1]. Our patient had elevated (32 times normal) urinary normetanephrine levels which confirmed the diagnosis.

The radiologic workup is usually performed after biochemical confirmation of catecholamine excess. Computed Tomography (CT) or Magnetic Resonance Imaging (MRI) is necessary to quickly locate PGLs [5]. MIBG (Meta-Iodo-Benzyl-Guanidine) scintigraphy has a specificity and sensitivity of 99 and 90%, respectively. It can detect multifocal, ectopic, and metastatic locations. However, it gives negative results in 10% of cases. Then other imaging techniques such as octreoscan and especially positron emission tomography using different tracers are required [6]. In our patient, the findings of abdominopelvic CT and MIBG scintigraphy were consistent with bilateral retroperitoneal paragangliomas.

Approximately 40% of PPGLs occur in the context of an autosomal inherited syndrome, making genetic testing essential [1]. Retroperitoneal forms are most often isolated. They are sometimes associated with other pathologies, notably Carney's

triad, multiple endocrine neoplasia type 2 and neurofibromatosis type 1 [3].

Therapeutic management usually consists of surgical excision, after careful drug preparation with alpha-adrenergic blockade and proper volume expansion [1]. Our patient benefited from a paraganglioma exeresis after a good medical preparation with alpha-blockers.

After surgery for functional PPGL, measurement of plasma and/or urinary methoxylated derivatives is recommended 2 to 6 weeks postoperatively.

Long-term follow-up for at least 10 years is essential due to the risk of local or distant recurrence (metastasis) and the risk of developing a new PPGL [1]. In our patient, the 24-hour urinary methoxylated derivatives were normal two weeks after surgery.

## Conclusion

Paragangliomas are rare tumours and their location in the retroperitoneal region is uncommon. However, it is important to make early diagnosis because of the life-threatening nature of the disease. Symptomatology is variable, non-specific and sometimes completely absent. The revelation of acute abdomen remains exceptional. The possibility of paragangliomas should always be considered when hypervascular masses are seen in certain locations of the body.

**Consent statement:** Written informed consent was obtained from the patient for publication of this Case report and any accompanying images.

## References


1. Cornu E, Belmihoub I, Burnichon N, Grataloup C, Zinzindohoué F, Baron S, et al. Phaeochromocytoma and paraganglioma. Rev Med Interne. 2019; 40: 733-741.
2. Ikram A, Rehman A. Paraganglioma. 2022. In Stat Pearls.
3. Fahmi Y, Elabbasi T, Khaiz D, Bensardi FZ, Hattabi K, et al. Retroperitoneal paraganglioma: A case report and review of the literature. Pan Afr Med J. 2015; 21: 298.
4. Laboureau S, Rohmer V. Pheochromocytoma et paraganglioma. EMC Endocrinologie-Nutrition. 2014; 11: 1-11
5. Yang Y, Wang G, Lu H, Liu Y, Ning S, et al. Haemorrhagic retroperitoneal paraganglioma initially manifesting as acute abdomen: a rare case report and literature review. BMC Surg. 2020; 20: 304.
6. Badaoui R, Delmas J, Dhahri A, Mahjoub Y, Fumery M, et al. Management of a retroperitoneal paraganglioma. Rev Med Interne. 2011; 32.