

Renal Hilar Paraganglioma - A Case Report

Rovena Bode¹, Medi Agolli², Rudin Domi³, Medi Alimehmeti⁴

^{1,2} Department of General Surgery, "Mother Teresa" University Hospital Center, Rruga e Dibrës, 1025 Tirana, Albania

² Department of Anesthesiology and Intensive Care, "Mother Teresa" University Hospital Center, Rruga e Dibrës, 1025 Tirana, Albania

³ Department of Histopathology, "Mother Teresa" University Hospital Center, Rruga e Dibrës, 1025 Tirana, Albania

Abstract: Objective: Paragangliomas are a type of neuroendocrine tumors derived from embryonal neural crest. When found in the adrenal gland they are called pheochromocytoma, when localized in the extraadrenal paraganglia they are called paraganglioma. Paragangliomas localized in the renal hilus are a rare entity. Methods: We are presenting a 30years old woman suffering from paroxysmal hypertension for more than a year and diagnosed with secretory paraganglioma. Ultrasound abdominal exam and CT scan confirmed a left renal hilar tumor. Urinary metanephrines and normetanephrines resulted elevated. Results: Laparotomy was performed and the removed mass was sent for histopathological exam. A paraganglioma was diagnosed. The operation resulted safe and effective. Now 5 years after operation the patient is healthy. Conclusions: Paragangliomas must be considered as a possible cause of hypertension in young people.

Keywords: pheochromocytoma, paraganglioma, chatecolamine metabolites, Zuckerkandel organ.

1. Introduction

Paragangliomas are neuroendocrine tumors derived from the embryonic neural crest. If localized in the adrenal gland they are called pheochromocytoma, if found extra-adrenal, arising along the sympathetic and parasympathetic paraganglia, they are called paraganglioma[1]-[2]. They are a category of chromaffin cell tumors, secreting chatecholamines in 50% - 60% of cases[2]- [3]. The extra-adrenal location accounts for 10-30% of cases, half of these arise from the organ of Zuckerkandel(fig.1) and the remainder from the retroperitoneum. Usually they are found in the abdomen, pelvis, thorax, neck and skullbase[4]. Very rare sites are the bladder, urethra, prostate, seminal vesicles and presacral area (Middledorpf tumor) [5]-[6].

Both paragangliomas and pheochromocytomas are characterized grossly by the brown appearance of the cut surface and microscopically by the presence of small nets of uniform polygonal chromaffin cells, surrounded by capillaries called Zellballen [7].

Men are affected more frequently than women, and most patients are between 30- 40 years [8]-[9]. Their incidence is 0.40-2.06 per million per year [10]-[11]. Characteristic symptoms are headaches, hypertension, palpitations, diaphoresis and sweating [2]. The above mentioned clinical table imposes a measurement of catecholamines and their metabolites (metanephrine and normetanephrine) in plasma and 24 hours urine samples.

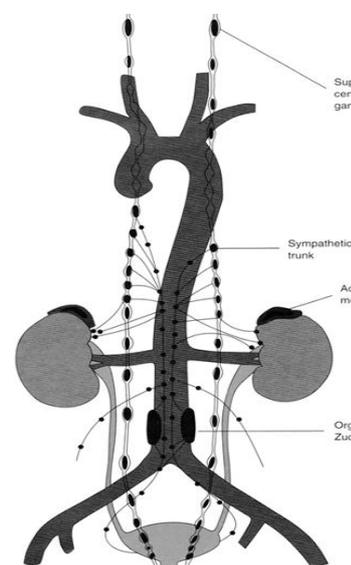


Figure 1: Organ of Zuckerkandel

2. Case Report

The patient is a 30-year-old woman. She presented in the general surgery department, with a 15 months history of paroxysmal hypertension, headache, pallor, diaphoresis and palpitations. The signs begin the last pregnancy, time when she was diagnosed by the abdominal ultrasound with a mass of 20*25 mm at the left renal hilum. The mass was grown and after a year the ultrasound revealed a 52*56 mm mass at the left kidney hilum.

The physical exam was non remarkable. No genetic predisposing factors in her familiar history. Complete cell count, blood urea nitrogen, creatinine and electrolyte levels of blood were all within normal limits. Urine analysis was unremarkable. Heart rate was 130 beats/ minute and fluctuation in blood pressure were noted. Elevated urinary metanephrines and urinary normetanephrines levels were recorded (respectively 1510ng/24 hours, 2814ng/24hours). Thyroid and calcitonin hormone levels were within normal

limits. Computed tomography of the abdomen demonstrated a left retroperitoneal tumor of 52*56 mm, extrarenal, located in contact with the left renal vein, laterally of aortal axis, with dense collateral circulation, no lymphnodes, no metastases (figure2, 3).



Figure 2: Ct-scan view of a hilar renal paraganglioma

Surgery was arranged 2 weeks after hypertension was medically treated by the endocrinologist. During surgery exploration, a 60*40 mm retroperitoneal tumor was found, encapsulated, which appeared soft and richly vascularised. It was localized between aorta and left kidney, laterally and inferiorly of cauda pancreatis, behind and inferiorly of spleen. The left adrenal gland was intact.



Figure 3: Ct-scan view of a hilar renal paraganglioma

Removal of the tumor was done and it was sent for histopathological examination (figure 4, 5).

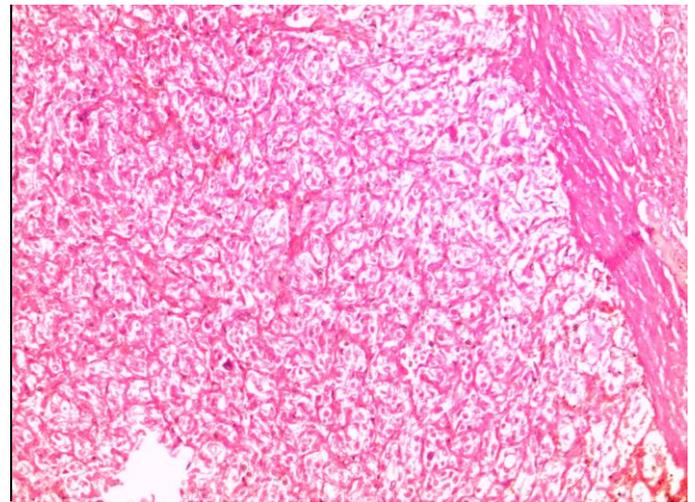


Figure 4: Histopathology view of the paraganglioma

The biopsy revealed a paraganglioma tumor. The patient had complete resolution, safely discharged from hospital. The 5 years follow up process revealed the patient is healthy.

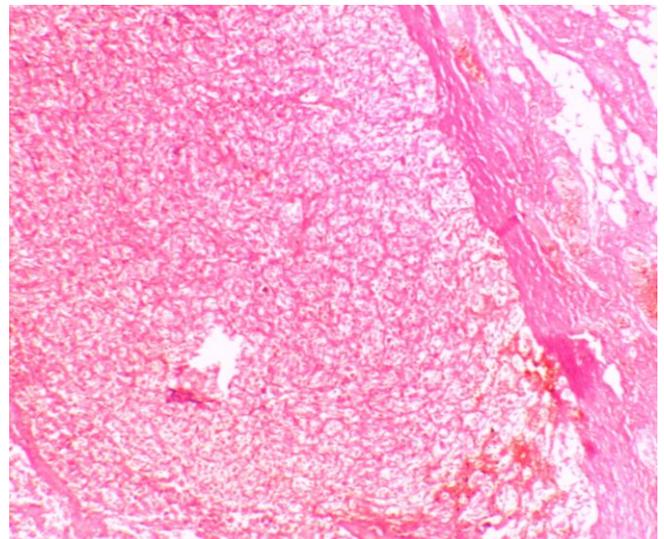


Figure 5: Histopathology view of the paraganglioma

3. Discussion

According to the World Health Organization classification of tumors, in 2004, paragangliomas are a type of neuroendocrine tumors derived from embryonic neural crest. They are mostly found in the adrenal glands (pheochromocytoma) and in the extra-adrenal paraganglia of the autonomic nervous system (paragangliomas)[1]-[4]. They usually occur sporadically, but they can be a feature of two disorders with an autosomal dominant pattern of inheritance: multiple endocrine neoplasia type 2 and 3 (MEN 2 and MEN 3) and von Hippel-Lindau disease. They may be part of Carney's triad associated with gastric leiomyosarcoma and pulmonary chondroma[12]- [13]. These tumors can be associated with clinical evidence of epinephrine secretion which makes them functioning tumors in 50-60% of cases [2]-[3], [7]. In fact the surgical manipulation of the neoplastic mass may produce a risky hypertensive crisis [2]-[3]. There are no histological criteria to distinguish between functioning and non functioning tumors[11]. But usually head and neck

paragangliomas, arising from parasympathetic paraganglions are nonsecretant. Histology and immunohistochemistry remain the gold standard for making the definitive diagnosis. Approximately 10% of them are malignant, nevertheless this cannot be determined on a biochemical or histological basis. It is the presence of local invasion, on gross or microscopic exam and metastasis presence that define malignancy [15]-[16]. Plasma free metanefrine and normetanefrine or urine metanefrine are the first tests to make the diagnosis of the disease [12]. Imaging studies such as CT-Scan and IRM are essentials before surgery for localization, size and evidence of metastasis. According to the NCCN guidelines the treatment of a paraganglioma is total resection when possible. If not, cytoreductive resection must be followed by radiation or systemic chemotherapy, depending on the extent of the metastases [17]. Paragangliomas are more likely to be malignant than pheochromocytomas (40% vs 10%) [2]-[15]. However evidence of metastasis is the only definitive criterion to label the tumor as malignant [2], [18]-[19].

4. Conclusion

In summary paragangliomas must be considered a possible cause of arterial hypertension in young people. Upon diagnosis, surgery is the only curative treatment. Adequate preoperative preparation and treatment, a multidisciplinary team (surgeon, endocrinologist, and anesthesiologist) can reduce morbidity and mortality.

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Author Profile



Rovena Bode Finished the study in Medicine University of Tirana, Albania in 2003. Graduated as General Surgeon in 2009 in the General Surgery Department, "Mother Tereza", University hospital Center, Tirana, Albania. Medical Emergency Training in Torino, Italy in 2010. During 2011 fellowship in Laparoscopic Surgery and breast surgery (oncology surgery) in Naples, Italy. Actually general surgeon in the Surgery Department "Mother Tereza" "University Hospital Center of Tirana, Albania.