Extra-adrenal paraganglioma of the retroperitonium: a case report and review of literature

Abstract

Paragangliomas are rare endocrine tumors that arise from paraganglia which are neuroendocrine non-excretory structures, derived from neural cells; they have a diffuse distribution related to the vessels and nerves of the branchial arches and to the ortho and parasympathetic systems. Paragangliomas can occur where the paraganglia exists. They can be found from the base of the skull to the pelvic floor, there seat of predilection is abdominal cavity, along the autonomic nervous system, usually within the para-aortic and perinephric spaces. The clinical presentation of this entity is marked generally with flank pain and hypertension which is generally difficult to manage, CT scan is the gold standard to predict the diagnosis and the treatment is surgical resection of the mass either in open or laparoscopic approach with a perfect monitoring of blood pressure; We present here a case of retroperitoneal paraganglioma treated in our institution.

Keywords: paragangliomas, perinephric spaces, pheochromocytoma, succinate dehydrogenase, metaiodobenzylguanidine, immunohistochemistry

Case report

A 33 year old female patient, with no medical or familial history. She was following up for arterial hypertension that was treated by beta blockers and calcium channel blockers. Her blood pressure remains difficult to control despite taking two antihypertensive agents. General examination revealed a patient in good general condition, apyretic with heart rate of 86 beats per minute, blood pressure of 170/10mmhg, average body mass index, cardiovascular examination did not show any abnormalities at the sites of auscultations or large vessels, the rest of the somatic examination were normal. An ultrasound with renal doppler has been demonstrated in the right renal hilum a well-limited rounded formation of tissue echostructure, hypoechogenic occurring in contact with the inferior vena cava and compressing the renal pedicle but without associated hemodynamic abnormality. Abdominal CT scan was performed (Figure 1) showing a retroperitoneal mass of the right renal hilum, enhanced intensely and early after injection of contrast agent indicative of its hyper-vascular character. Kidney and adrenal glands were free of abnormalities.

In view of the findings of the imaging and this clinical presentation, a dosage of methoxylated catecholamine derivatives in the 24-hour urine was performed, demonstrating an elevation of normetanephrine: 42.74μmol/24h (N: 0.40 to 2.10) and metanephrine: 1.18μmol/24h (N: 0.20 to 1.00). Our patient has undergone a surgical intervention with the aim of complete resection of this mass. Perioperative monitoring was performed by ETCO2, SpO2, ECG, temperature, diuresis, arterial pressure by arterial line and central venous access.

We found an Oval mass located in front of the renal pelvis and the proximal lumbar ureter and outside the IVC displacing the renal pedicle up, the right adrenal were intact. The preoperative evaluation was marked by the occurrence of a hypertensive peak with PAS at 19mmhg treated with a bolus of nicardipine. Macroscopic examination of the surgical specimen (Figure2) found an oval mass of tissue consistency well limited by a measuring capsule with abundant blood supply at measuring 5×3, 5×3 cm.
The histological study concluded in a morphological aspect in favor of a paraganglioma (Figure 3). The postoperative course was uneventful. Our patient left the hospital five days after surgery. She was seen one month later with normal values of blood pressure and an I-123-MIBG-scintigraphy was performed but didn’t show any abnormalities. It has been planned to send her for genetic counseling in search of a predisposing genetic abnormality.

Discussion

Also called extra adrenal pheochromocytoma, since they’re developed from the extra-adrenal chromaffins embryonic residues. Paragangliomas are rare endocrine tumors that arise from paranglia which are neuroendocrine non-excretory structures, derived from neural cells; they have a diffuse distribution related to the vessels and nerves of the branchial arches and to the motor and para-sympathetic systems. Paragangliomas can occur where the paranglia exists. They can be found from the base of the skull to the pelvic floor there seat of predilection is the the abdominal cavity, along the autonomic nervous system, usually within the para-aortic and peri-nephric spaces. Pheochromocytomas are extremely rare tumors with an incidence of 2 to 8 per million. It believed that 10% of all pheochromocytomas are at extra-adrenal sites. But there are reports of an incidence as high as 22% in adults opposed to this traditional teaching. Retroperitoneal paragangliomas mainly affect adults who are in the fourth or fifth decade of life, and they have no sex predilection. They occur either sporadically or as part of a hereditary syndrome. At least one-third of all patients with PPGILs have disease-causing germline mutations. Autosomal dominant mutations in succinate dehydrogenase (SDH), particularly in the SDHB subunit, are the most frequent germline abnormalities. It has been planned to send her for genetic counseling in search of a predisposing genetic abnormality.

These tumors are identical histologically to pheochromocytomas. The extra-adrenal paraganglioma system is composed predominantly of chief cells, arranged in compact nests (zellballen), and peripheral to these nests there is sustentacular cells within a highly vascularized stroma. It has been shown that all paraganglia store catecholamines in dense core granules of the chief cell and, therefore, theoretically possess the potential for functional secretory status. Thus we distinguish functional and non-functional Paragangliomas. Most retroperitoneal paragangliomas present with symptoms related to catecholamine excess dominated by Paroxystic or permanent hypertension as well as the classic triad of Menard combining headaches, sweating and palpitations. 14% of patients have either atypical symptoms or are asymptomatic. And be revealed until an advanced stage of the disease or by serious complication. The case of Kwok Kay Yau et al. in which they report a spontaneous rupture of nonfunctioning retroperitoneal paraganglioma that presented as acute abdomen dramatically illustrates this point. Biological diagnosis is based on a biochemical testing by the measurements of catecholamines and their metabolites. Plasma free metanephrines or urinary fractionated metanephrines are superior to other tests of catecholamine excess for diagnosis of paragangliomas and have more than 90% sensitivity for Paraganglioma. Plasma and urine catecholamines have frequent false-positive results. Careful attention should be given to medications which interfere with the testing of plasma metanephrines such as tricyclic antidepressants, monoamine oxidase inhibitors, and certain beta-adrenergic and alpha-adrenergic blockers selective serotonin reuptake inhibitors acetaminophen. These medications should be stopped for 10 to 14 days before testing if possible. Their topographic diagnosis is based on ultrasound, computed tomography and Metabolimobenzylguanidine scintigraphy which has often been used as an imaging modality in the diagnosis of neuroendocrine tumors. Currently magnetic resonance imaging (MRI) is the most reliable examination to detect paraganglioma and even small metastases. In a review of 236 cases of paraganglioma, metaboliod benzylguanidine (MIBG) scintigraphic study was the least sensitive imaging study compared to CT and MRI. Although lacking in sensitivity, MIBG scan is highly specific and may be the only positive imaging test in some patients. Complete surgical excision is the mainstay of treatment for Paraganglioma. This has been confirmed by the outcome of 22 cases of extra-adrenal retroperitoneal paraganglioma from Memorial Sloan-Kettering Cancer Center (MSKCC), where the five-year survival for tumors not resected was only 19% compared with 75% after complete resection. As with pheochromocytomas paragangliomas require careful preoperative preparation to prevent the significant risks of mortality and morbidity due to paroxysmal catecholamine releases.

Due to the extreme anatomical, biochemical, genetic, biological, and clinical variability and heterogeneity of paraganglioma. There are currently no firm guidelines for the management of patients with paraganglioma. Only observational data and recommendations based on the opinions of experts in the field are at present available. Patients with a biochemically positive Paraganglioma must be treated with medical blockade before any surgery or procedure to avoid the effects of released catecholamines. α-Adrenoceptor antagonists are the drugs of choice and the most widely used. The current surgical techniques and medical management have significantly decreased the mortality rate. Both classic and robot-assisted laparoscopic approach have proved their feasibility and safety for resection of retroperitoneal paraganglioma with a good results despite the diversity of the lesion sites and the complicated relationship with great vessels. The prognosis of paragangliomas is very difficult to predict due to their heterogeneity. In addition to the deficiency of data from the literature on their natural history and outcome after resection, the criteria for malignancy are not well established. A paraganglioma is defined, by the World Health Organization, as malignant only when it presents with one or more distant metastatic localizations where chromaffin tissue is not usually present. Prognostic systems predictive of malignancy have been developed but are still not validated. Because of malignant potential and higher recurrence rate in paragangliomas, lifelong follow-up is always recommended.

Conclusion

Functional PGL is a rare cause of secondary arterial hypertension, providing great cardiovascular morbidity and mortality. It must be considered in any arterial hypertension in young patients. Histological diagnosis is made easy with immunohistochemistry techniques. Preoperative pharmacologic preparation, attentive intraoperative monitoring, and aggressive surgical therapy have important roles in achieving successful outcomes. Recent reports suggest that a laparoscopic approach can safely remove these tumors. The identification of predisposition genes has modified the management of these tumors. Currently screening and monitoring of patients and their family members may help in the near future for establishing personalized medical management for affected patients.

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Conflict of interest

Authors declare there is no conflict of interest in publishing the article.

References