

A Rare Case of Retroperitoneal Paraganglioma Expanded Intraoperitoneally

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Abstract: Paraganglioma is a rare extra-adrenal tumor of the autonomic nervous system which is usually occur in the head and neck region but rare in the retro peritoneum.

We will report a case of secreting paraganglioma originated from retro peritoneum expanded into the peritoneum.

The mass being unresectable, the patient was treated by radiotherapy.

Date of Submission: 20-03-2019

Date of acceptance: 06-04-2019

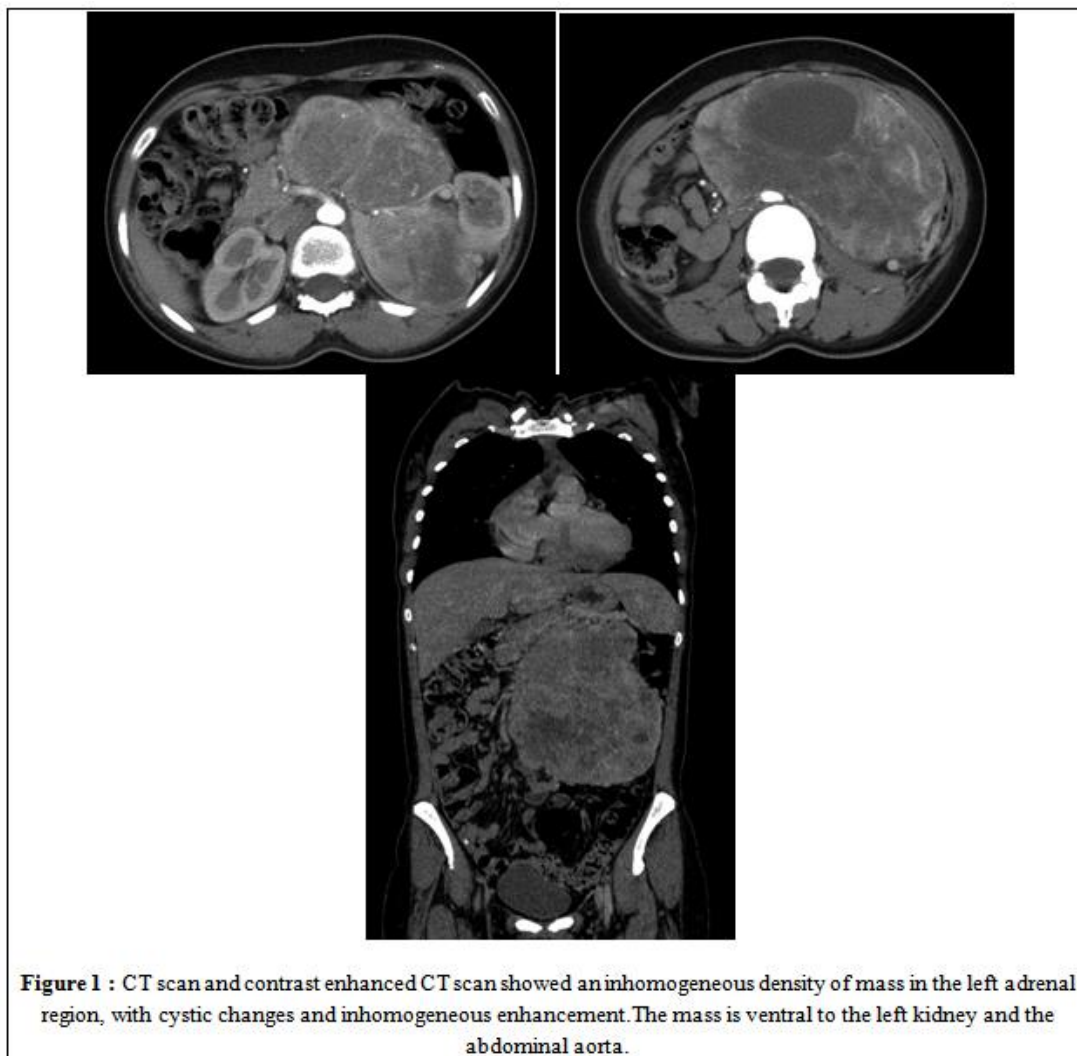
I. Introduction

The paraganglionic system is composed of neural crest cells, which are found in the adrenal medulla, parasympathetic ganglia, and chemoreceptors. Tumors that arise from the chromaffin cells of adrenal medulla are called pheochromocytomas, and those that arise in an extra adrenal location (10%) are referred to as paragangliomas [1, 2].

II. Case Report

Herein, we will present the case of a thirty four year woman who had an increasing degree of pain and coving in the left flank. She had hypertension and palpitations for three months before consulting. On clinical examination, there is a sensitive firm mass occupying the left lumbar fossa and left flank measuring approximately 15 cm with a blood pressure of 150/70 mmhg.

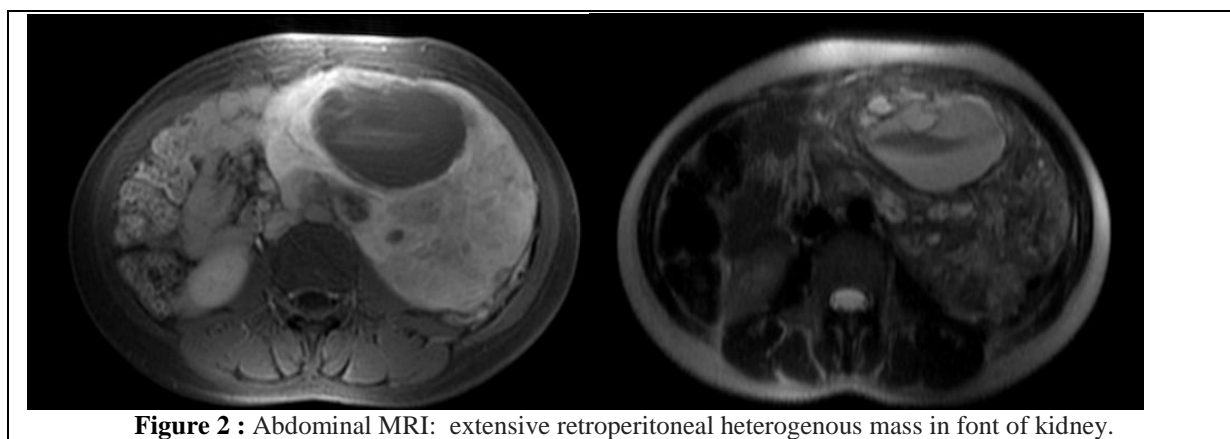
CT scan and MRI have been realized. Computed tomography objective to the presence of a voluminous fleshy and liquid retroperitoneal mass, lobulated, in front of the left kidney with a peritoneal extension. It measures 17 x 13 x 12 cm (figure 1)



The tumor drives back the spleen and the left kidney, engages the renal pedicle and compresses the proximal ureter with mild pyelo-caliceal ureteric dilatation.

Inside it passes the median line, it pushes back the tail on the right, the head of the pancreas and the upper mesenteric vessels without sign of invasion.

Abdominal MRI : heterogeneous mass was found in the front of left kidney in the retro peritoneum. It showed low signal on T1, intermediate and high signal were shown on T2 WI and T2WIFS. High signal intensity was shown on DWI.(figure 2)



There was a significantly enhanced signal on contrast phase.

The biological assessment showed a hemoglobin to 8.8 g/dl, the 24 hour urine test showed that norepinephrine level was high.

A true cut biopsy was performed and sent for frozen as well as permanent section analysis.

Histological and immune-histochemical appearance was consistent with paraganglioma.

He has demonstrated a tumor proliferation arranged in small nests and layers supported by a vascularization giving him an endocrinoid architecture.

Tumor cells intensely expressed chromogranin and synaptophysin and don't expressed CKAE1/AE3.

Tumor proliferation index KI67 was estimated at 5%.

The tumor was unresectable, the patient received radiotherapy with total dose of 50 GY : 2Gy per day dor 25 sessions (end of t irradiation April sixth 2017).

CT scan examinations were performed afterwards. The last control imaging performed on January 16 , 2018 notes a regression in size of the tumor process measuring 12.5 x 8 x 14.5 cm versus 17 x 13 x 12 cm (figure 3) .



Figure 3 : Regression in size of the retro-peritoneal tumor currently measuring 17 X 13 X 12 mm versus 12.5 x 8 x 14.5 cm

III. Discussion

Primary retroperitoneal neoplasms are rare benign and malignant mesenchymal tumors that arise in the retro peritoneum [3].

Paraganglioma are extra adrenal pheochromocytomas that arise from chromaffin cells in the sympathetic or parasympathetic neural paraganglia [4]. They are account for 1–3% of retroperitoneal tumors.

Retroperitoneal ganglioma can be divided into functional and non-functional (clinically silent). Functional tumours are often associated with hypertension, tachycardia, headache and diaphoresis [5]. However, non-functional paragangliomas can be completely clinically silent. Functional paragangliomas secrete norepinephrine and normetanephrine account for 30-60% of the tumors [6].

The diagnosis is usually established with high urine catecholamine metabolites , VMA and metanephrine levels [7].

On CT, retroperitoneal paraganglioma appears as a hypervascular mass. Areas of intralesional hemorrhage and necrosis can be frequently seen as the tumor enlarges [8].Punctate calcification is seen in 15 % of cases, and a fluid –fluid level can be seen that is due to hemorrhage [1].

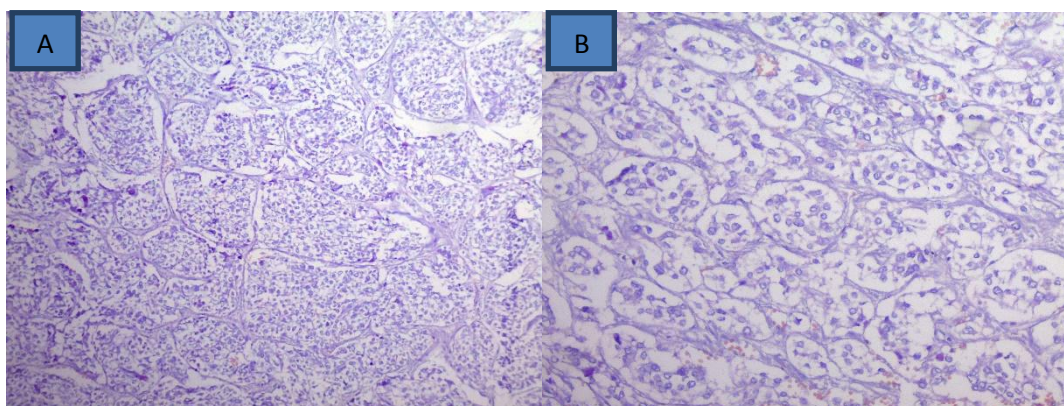
Paragangliomas are usually decribed on MRI as masses having characteristic high signal intensity or a “light bulb bright” signal on T2WI with the use of Fat suppression and hypointensity on T1WI [9].

They also have a strong initial signal during the arterial phase [10]. It was the case of our patient. The tumor is strongly enhanced at arterial phase.

Metastasis of paraganglioma of the retro peritoneum is usually to lung, lymph nodes, liver, bones or the spleen [2]. Our patient did not have distant metastasis.

Radionuclide imaging performed after administration of meta-iodobenzylguanidine (MIBG) shows high uptake in paragangliomas and is a sensitive technique for localizing these lesions [1].Recently, the utility of PET has been investigated. There are several radiotracers that can be used with PET, although it has a high sensitivity for metastatic disease. [11].However , specific diagnosis for the retroperitoneal mass still relies on post-biopsy histological diagnosis. Chromogranin A and synaptophysin are the most common neuropeptides synthesized in endocrine cells and can be used for immunohistochemical analysis of paragangliomas along with

other protein markers such as neuron specific enolase and vimentin [2]. Histological analysis confirmed the diagnosis in our patient (figure 4).



- A. Microphotography showing a nested pattern, made of nests (Zellballen) separated by anastomosing bands. (HE, 100X)
- B. Microphotography at higher magnification, showing that tumor cells are round to oval with abundant granular eosinophilic or basophilic cytoplasm. (HE, 200X)

Surgery is the preferred treatment of paraganglioma. Phenoxybenzamine should be orally administered 6 weeks before surgery to relieve symptoms [12]. Radionuclide therapy and systemic chemotherapy had been reportedly used for malignant paraganglioma with distant metastasis [13]. Radiation therapy has been advocated for patients who cannot undergo surgery or for unresectable tumors [13]. This is the case for our patient, in whom the tumor has intimate vascular reports.

IV. Conclusion

In conclusion, retroperitoneal paraganglioma are rare tumors, mostly benign with good prognosis, but can be locally invasive and metastasize as well. The diagnosis of paraganglioma depends on the clinical history, the presence of urinary or circulating findings, and tissue histopathology. Surgical excision remains the mainstay of treatment, although advanced disease and prominent vascularity can at times make excision difficult or impossible.

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