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A Case of Clinically Silent Giant Right Pheochromocytoma

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Abstract **Case Report**

Patients with pheochromocytoma are typically symptomatic. Hypertension is the most common sign of pheochromocytoma. Clinically silent giant pheochromocytoma is a rare adrenal disease; complete resection is the only curative treatment. Due to the serious surgical risk, successful resection is very difficult. We report a case of 70-year-old- female with giant pheochromocytoma, which was successfully resected. There were no intraoperative and postoperative complications. We analyze and report our experience.

Keywords: Pheochromocytoma, retroperitoneal mass.

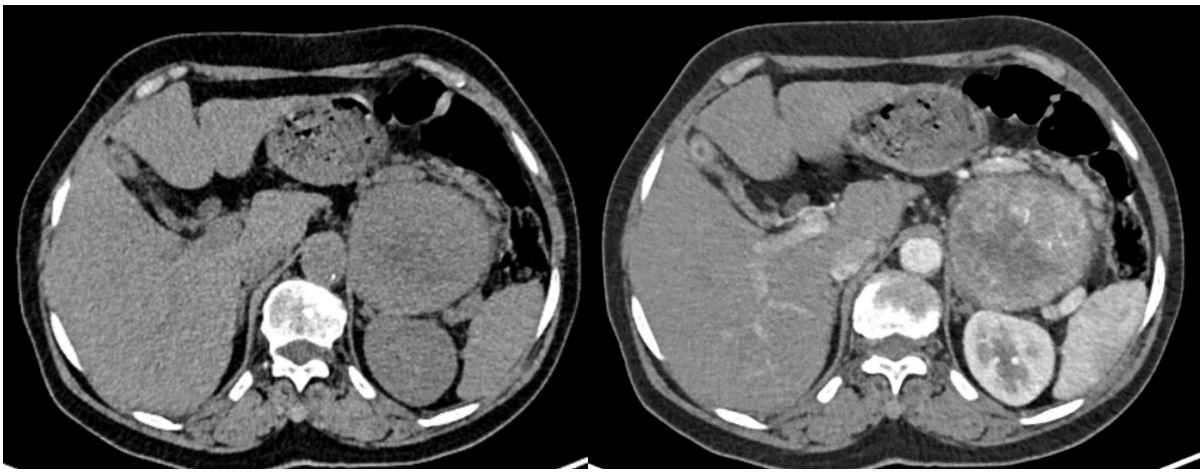
INTRODUCTION

Pheochromocytomas are catecholamine producing tumors which arise from chromaffin cells within the adrenal medulla. Silent pheochromocytomas are rare entities that do not present with the classical symptoms commonly seen in catecholamine-secreting tumors.

CASE REPORT

We report a case of 70-year-old- female patient who presented with left sided abdominal pain and discomfort for 6 months.

A preoperative Computed tomography (CT) scan showed a voluminous retroperitoneal left adrenal mass 90 X112X 153 mm hypodense in spontaneous contrast, with moderate and heterogeneous enhancement, with central necrosis zone (Figure 1).



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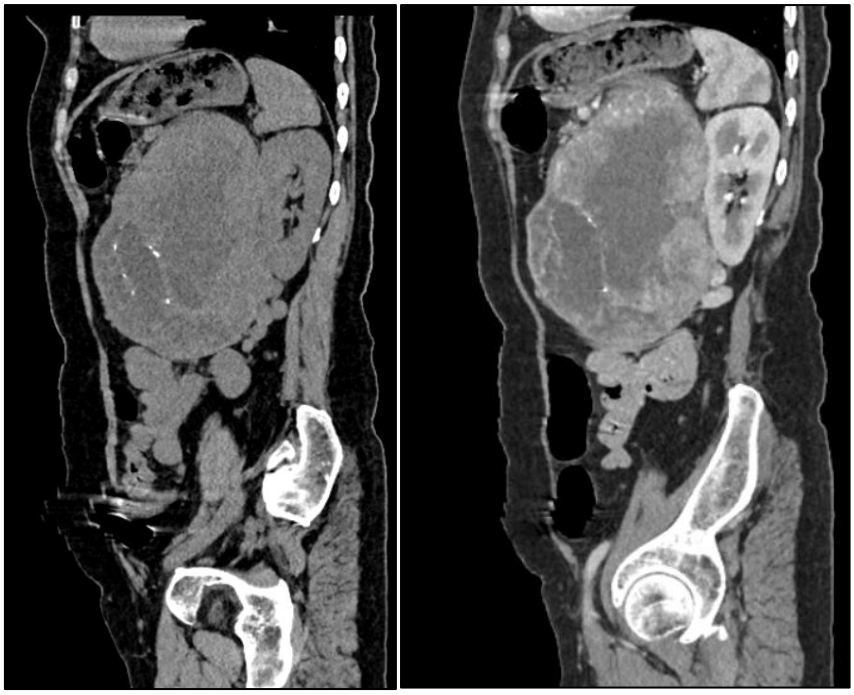

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Fig. 1: Axial and sagittal view of Non-enhanced CT (A, B) Axial and sagittal view of enhanced CT demonstrates: Voluminous retroperitoneal left adrenal mass 90 X112X 153 mm hypodense in spontaneous contrast, with moderate and heterogeneous enhancement, with central necrosis zone

The right adrenal gland was normal and there were no finding of distant metastatic. The urinary catecholamines were very elevated:

- **Normetanephrine:** 29.31 mg/ 24 h (0.07- 0.46)
- **Métanéphrine:** 24.03 mg/ 24h (0.04-0.30)
- **Creatinurie:** 0,69 g/ 24H (0.95-1.60)

The patient underwent a laparotomy adrenalectomy after failed laparoscopy resection. The mass weighed 1137g (Figure 2).

Histopathologic and immunohistochemistric evaluation confirmed the diagnosis of pheochromocytoma confirmed with a PASS score of 8.

The immunohistochemistry found: CgA(+), Synaptophysin (+), melanA(−), PS100.

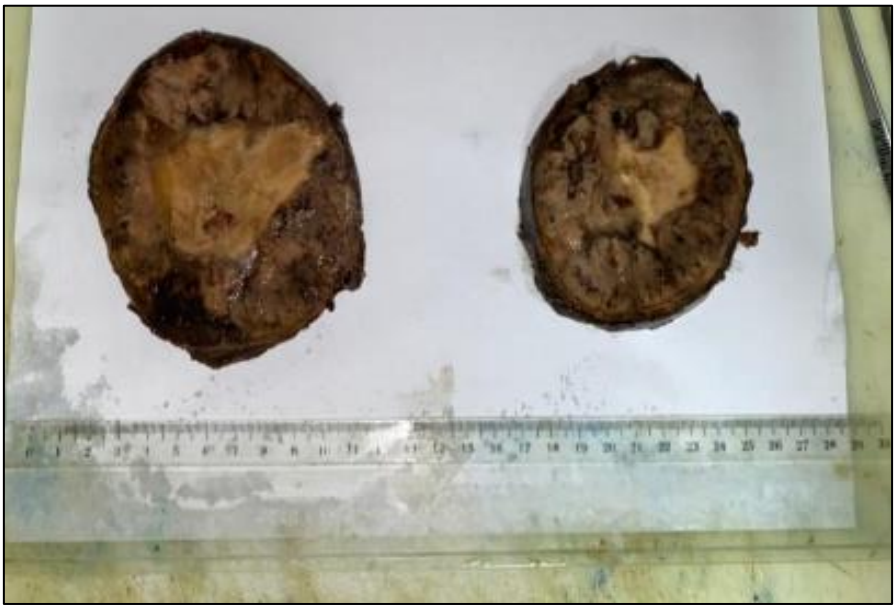

Fig. 2: The tumour measured 15 cm × 12 cm × 6 cm and weighed 1137 g. The cut surface was tan with extensive haemorrhagic and focal necrotic areas

The patient is on long term follow up. After six months of follow-up, our patient was completely asymptomatic and the urinary catecholamines were negative.

DISCUSSION

Patients with pheochromocytoma are typically symptomatic. Hypertension is the most common sign of pheochromocytoma [1, 2]. Giant pheochromocytoma is by definition tumors more than 7 cm and is rare.

Of all the adrenal pheochromocytoma, 20% to 30% of them are asymptomatic; they are called clinically silent pheochromocytoma. They present with vague abdominal symptoms .On the basis of symptoms and signs, the patient should be defined as clinically silent pheochromocytoma [3].

It is also important to note that it is not possible to distinguish malignant from benign pheochromocytomas merely on the direct appearance of the mass. Rather, the distinction is made on demonstrating evidence of direct tumor invasion into adjacent organs/structures or the presence of metastases [4].

On ultrasound Pheochromocytomas can have a variable appearance ranging from solid to mixed cystic and solid to cystic [5].

Computed tomography (CT) scan is the gold standard in the diagnosis of giant pheochromocytoma.

CT is the first imaging modality to be used, with an overall sensitivity of 89%. This is on account of 98% of tumors being located within the abdomen and 90% limited to the adrenal glands [6], and demonstrates usually large, heterogeneous masses with areas of necrosis and cystic change and they typically enhance avidly [7] may wash out similar to an adrenal adenoma, but they tend to have a greater enhancement in an arterial or portal venous contrast phase [4].

It should be noted that suspected cases of pheochromocytomas have been historically managed as a contraindication for iodinated contrast administration, as it could theoretically precipitate a hypertensive crisis. However, studies have shown no evidence to support this theory and nowadays most radiology non-ionic iodinated contrast media guidelines do not place pheochromocytomas as a contraindication to iodinated contrast administration [8, 9].

MRI is the most sensitive modality for the identification of pheochromocytomas and is particularly useful in cases of extra-adrenal location. The overall sensitivity is said to be 98% [6].

T1W sequence demonstrates: Slightly hypointense to the remainder of the adrenal**,** if necrotic and/or hemorrhagic then the signal will be more heterogeneous.

T2 sequence demonstrates: Some are markedly hyperintense (lightbulb sign): ~1/3 do not have marked T2 hyperintensity [10] areas of necrosis, hemorrhage and calcification will alter the signal.

In/out-of-phase sequence shows: No signal loss on out-of-phase imaging (pheochromocytomas rarely contain a significant amount of intracellular lipid) [11].

T1 post contraste (Gadolinium) images demonstrate: Heterogeneous enhancement, enhancement is prolonged, persisting for as long as 50 minutes [4].

Over 70% of tumors express somatostatin receptors. Imaging is obtained 4 hours (+/- 24/48 hours) after an intravenous infusion [12].

Octreotide is usually labeled with either 111In-DTPA (Octreoscan) or (less commonly) 123I-Tyr3-DTPA [12].

I-123 MIBG uptake in an adrenal nodule is strong supporting evidence for a pheochromocytoma. Overall sensitivity is ~80% 6. However, as many neuroendocrine tumors demonstrate uptake with MIBG, it is not as specific for pheochromocytoma outside of the adrenal.

18F-DOPA PET is thought to be highly sensitive according to initial results [13]. Gallium-68 DOTATATE PET-CT imaging has also been advocated due to their higher lesion to background tissue contrast and high specificity for pheochromocytoma [14].

18F-FDOPA (6-[18F]-L-fluoro-L-3, 4-dihydroxyphenylalanine)-based PET-CT imaging can also be a useful tool in diagnosis [15].

The majority of pheochromocytomas including the giant category are benign. Biologically aggressive tumors have been found to generally have a PASS score ≥4. However, this is not definitive, and is only suggestive of malignancy. Histopathological diagnosis of malignant pheochromocytoma is made if ectopic chromaffin cells are detected in the extra-adrenal sites [3, 4].

Once the adrenal neoplasm is diagnosed, resection is the only curative treatment. Although some reports have concluded that laparoscopic adrenalectomy is safe, effective, and minimally invasive, it is considered a gold standard in the surgical management of small benign adrenal tumours. In our case, considering the large size of the tumour (>10 cm), small surgical space and possible malignancy with local invasion, a traditional open surgery was performed after conversion from coelioscopy [16, 17].

Wang Dong and colleagues reported that selective arterial embolization before surgery can

significantly decrease blood loss. However, due to the extensive abnormal vascularization with multiple arterio-venous communications, the authors found it difficult to complete arterial embolization. A multidisciplinary approach is advocated for successful management of phaeochromocytoma [18].

CONCLUSION

This case brings to the attention of clinicians the need to have a high index of suspicion of a giant pheochromocytoma in a patient presenting with vague abdominal symptoms whose CT scan shows a large retroperitoneal tumor, even in the absence of clinical symptoms.

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