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BENIGN GIANT PHEOCHROMOCYTOMA -A CASE REPORT OF A BIOLOGICALLY AGGRESSIVE TUMOR

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ABSTRACT

Pheochromocytomas are rare benign tumors located in the adrenal medulla. Giant Pheochromocytomas are generally classified as those with maximal diameters greater than 10 cm. The most common clinical feature is hypertension and only 50% of the patients present symptoms compatible with this pathology. Here we describe the case of a 53-year-old woman with biologically aggressive Pheochromocytoma. Also among one of the largest pheochromocytoma reported in India, as per the available indexed literature.

INTRODUCTION

Pheochromocytoma accounts for less than 0.1% of the hypertensive population. Hypertension is one of the most common manifestations of pheochromocytoma and can be persistent or paroxysmal. Diagnosis is confirmed by raised plasma metanephrines or normetanephrine. The triad including episodic headache, palpitation, and sweating occurs in 24% of pheochromocytoma patients [1][2]. We present a case of a 53-year-old woman with one of the largest biologically aggressive Pheochromocytoma reported in India, as per the available indexed literature which forms the basis of this communication.

Case Report

A 53-year-old female patient presented with a complaint of left hypochondrium pain for 1 month with the complaint of headache and palpitation. Known diabetes mellitus and hypertension. USG abdomen and pelvis showed a solid mass lesion 11.2x9.4x8 cm abutting the upper pole of the left kidney. CECT abdomen and pelvis showed a mass in the left suprarenal gland. adrenocortical carcinoma, metastasis. PET-CT showed a large soft tissue mass in the left suprarenal region 9x10x8.7 cm. Plasma metanephrine 80 pg/ml, plasma nor-metanephrine >7200 pg/ml, serum ACTH- 42pg/ml, serum cortisol-14.7 mcg/dl, DHEA-85mg/dl. With adequate preoperative preparation left open adrenalectomy was done. 13cm x9 cm x8 cm left suprarenal mass resected uneventfully. The histopathology evaluation revealed evidence of left pheochromocytoma with intravascular invasion and high cellularity which indicates PASS >4. A Pheochromocytoma of the Adrenal gland Scaled Score (PASS) weighted for the specific histologic features can be used to separate tumors with a potential for a biologically aggressive behavior (PASS > or =4) from tumors that behave in a benign fashion (PASS <4). The pathologic features that are incorporated into the PASS correctly identified tumors with a more aggressive biologic behavior. Post-surgery after 6 months, no significant abnormality was noted on ultrasonography, and plasma normetanephrine dropped to 130 pg/ml.

Table 1. Pheochromocytoma of the Adrenal Gland Scaled Score (PASS)

Characteristic	Score
Large nests or diffuse growth	2
Central or confluent tumor necrosis	2
High cellularity	2
Cellular monotony	2
Tumor cell spindling	2
Mitotic figures >3/10 HPF	2
Atypical mitotic figure(s)	2
Extension into adipose tissue	2
Vascular invasion	1
Capsular invasion	1
Profound nuclear pleomorphism	1
Nuclear hyperchromasia	1
Total	20

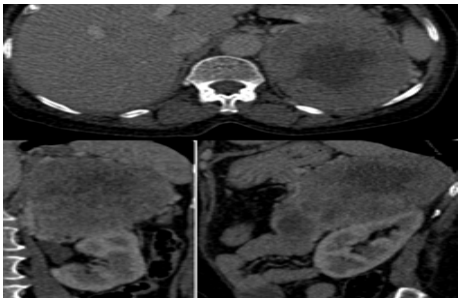

Fig. 1: CT scan showing left adrenal mass

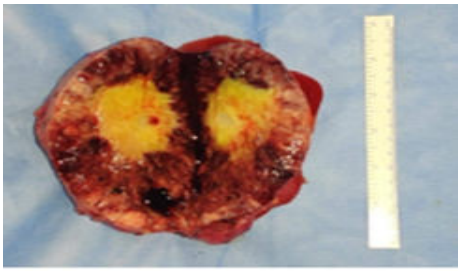

Figure. 2: Left Adrenalectomy specimen

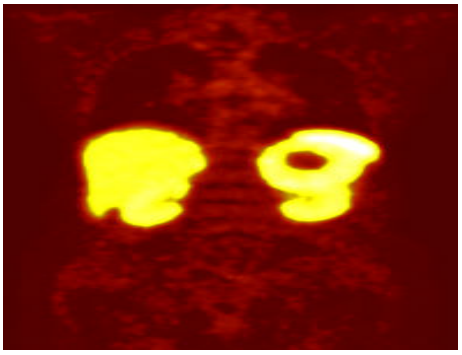

Fig 3- PET/CT showing a large mass in the left suprarenal region

DISCUSSION

Pheochromocytomas are most common in the fourth to fifth decade but can be seen at any age. They are common in both sexes[3]. The clinical presentation varies with 4% of adrenal masses incidentally found being known to be pheochromocytomas and the rest are benign adenomas. The diagnosis of pheochromocytoma requires both proof of excessive release of catecholamines and anatomical documentation of the tumor. Surgery is the treatment of choice for these tumors and cures 90% of patients. The preoperative preparation is essential to reduce the perioperative morbidity and mortality in these patients[4] . Post-surgery hypotension can occur in 30-70% along with hypoglycemia. Hence blood pressure and random blood sugar are monitored post-surgery[5].

CONCLUSION

Pheochromocytomas are rare neuroendocrine tumors responsible for less than 1% of hypertensive cases. Only 50% of individuals will present symptoms with this tumor. However, surgery is curative for pheochromocytomas like in our patient. In our first 6-month follow-up, our patient remained disease-free, but biological aggressiveness on HPE warrants long-term surveillance.

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