Case Report

UNIQUE CASE OF BILATERAL ADRENAL PHEOCHROMOCYTOMAS: DIAGNOSTIC APPROACH

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ABSTRACT

Pheochromocytoma is a neuroendocrine tumor arising from the adrenal medulla. Paragangliomas also originate from neural crest tissue but outside the adrenal gland. The presence of Bilateral adrenal masses is a unique and reportable event. Bilaterality is a treatment dilemma for a clinician whether to operate or not, keeping in mind that it can render patients with life threatening complications, perioperative mortality, and the potential burden of life long medications in some cases. We present a unique case of 28 years old female who presented with left lumbar pain, occasional headaches in her twenties followed by spells of uncontrolled hypertension during pregnancy, referred in post-partum period following a successful pregnancy to urology with suspicion of bilateral adrenal masses ending up with histopathology and immunocytochemistry confirming the diagnosis of Bilateral Adrenal Pheochromocytoma

Key Words: Pheochromocytoma, Adrenalectomy, Pregnancy, Adrenal medulla

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INTRODUCTION

Pheochromocytomas and Paragangliomas are rare tumors of neural crest origin.[1,2] Some of them present with symptoms while most are diagnosed as incidental findings on imaging done for other purposes. Symptoms may include episodic headaches, sweating, tachycardia, flushing, and paroxysms of hypertension.[3] Due to overlaps of symptoms with various other medical conditions there is often seen a delay in patient presentation and correct diagnosis.[3] Diagnostic workup includes measurements of urinary catecholamines secreted by the tumor and imaging including ultrasound, CT scan, MRI and MIBG scans where suited.

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Pheochromocytomas being a rare diagnosis may mimic Renal Cell Carcinoma.[4] It is a diagnostic challenge for the clinician and as well as a difficult pathological diagnosis for the histopathologist sometimes requiring re-reporting of slides by a second pathologist or a second review by outsourcing to confirm the diagnosis. Pheochromocytomas due to the release of Catecholamines during surgical manipulation for resection cause unpredictable fluctuations in blood pressure during the surgery as well as in the immediate post operative period requiring a dedicated anesthesia team and ICU care. Adrenalectomy in such cases should be performed by the senior surgeon as the surgery requires a high set of surgical skills. Surgical resection is further made difficult by the rich blood supply and the potential threat of bleeding with such fluctuating per operative blood pressures.[5]

We present a unique case of 28 years old female presenting with bilateral adrenal masses who underwent bilateral

adrenalectomies with histopathology and further immunostaining confirming the diagnosis of Bilateral Pheochromocytomas.

CASE REPORT

A 28 years old female was referred to us by the gynecologist after a successful pregnancy with spells of uncontrolled hypertension starting from 7-8 weeks of pregnancy and continuing throughout the antenatal period. She underwent an Ultrasound abdomen later during the pregnancy showing evidence of hypoechoic lesions bilaterally in the Suprarenal region with no pathology seen in the kidneys bilaterally. After the successful pregnancy, further workup was advised including a CT abdomen and pelvis with contrast which showed bilateral adrenal masses of more than 5cm in size each showing intense enhancement and nonenhancing areas of necrosis. Nodular calcification was noted on the left side. The fat planes with kidneys were clear. (Figure 1) She had her urinary catecholamines levels done in which Urinary metanephrines levels of 134.6 pg/ml, normetanephrine levels of 800 pg/ml and dopamine levels of 93.4 pg/ml were seen, further supporting the diagnosis of Bilaterally functional or either one functional pheochromocytomas. After meticulous pre operative preparations including blood pressure monitoring she had her Left Open adrenalectomy done with uneventful post operative recovery except for minor fluctuations in blood pressure. This did not require any need for blood transfusions.

Upon histopathology of this left adrenal mass, the Gross cut section showed a yellowish brown lesion with a white gelatinous central nodule. Histopathological examination showed cells arranged in nested and solid patterns with abundant eosinophilic granular cytoplasm with nuclei having salt and pepper chromatin. Typical Zellballen patterns of polygonal cells were also seen. To confirm this rare diagnosis second review of histopathology was done by another pathologist, both suggesting the same. Further immunohistochemical staining of the same slides showed positive results for Chromogranin and negative staining for CK, PAX8, CAIX, SMA, HMB45 and Inhibin. Based upon the above the histopathological diagnosis was confirmed as Pheochromocytoma. (Figure 2, Figure 3)

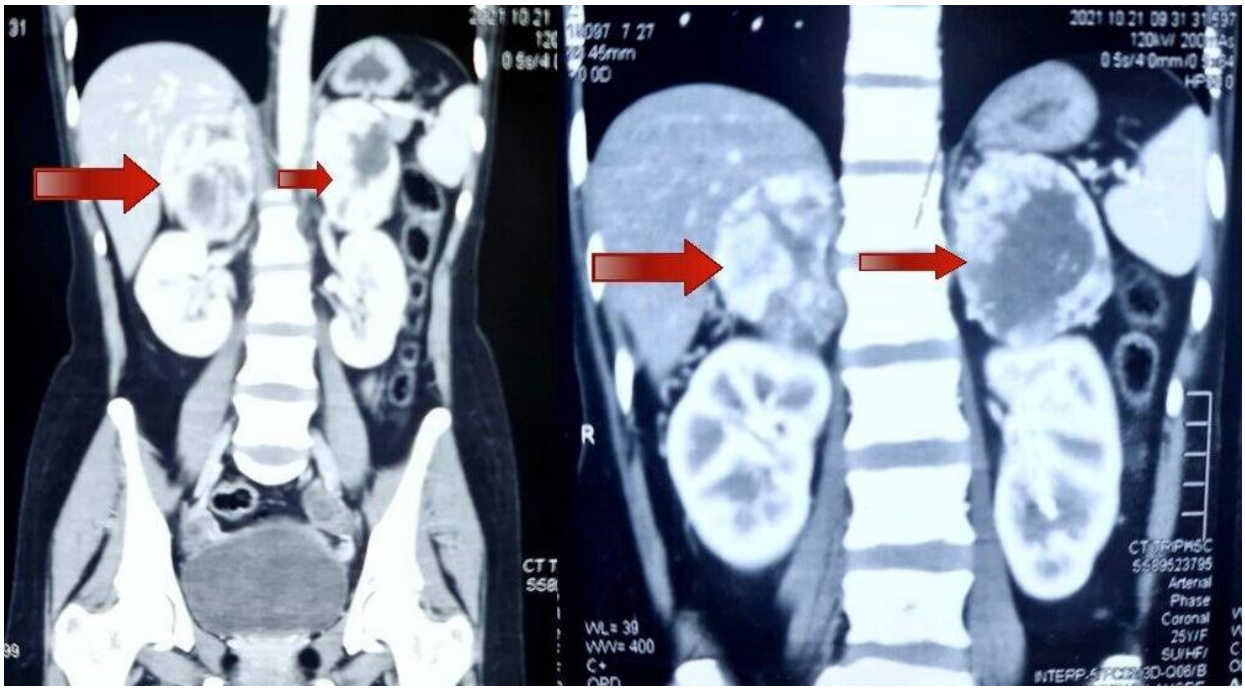

Figure-1: CT Abdomen and Pelvis showing Bilateral Adrenal Masses

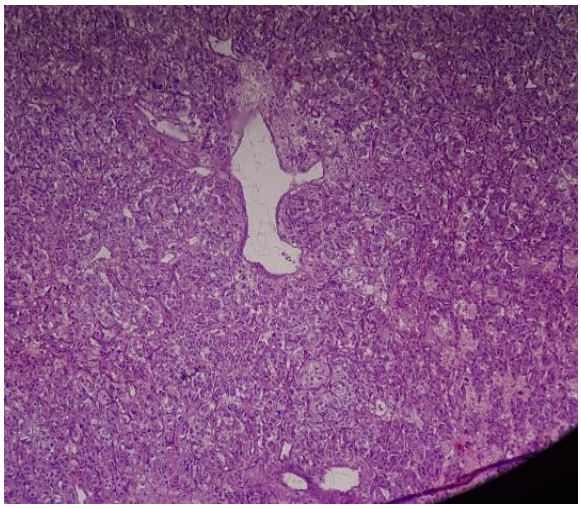

Figure-2: Left adrenal mass low power view (4x) showing packeting of cells zell Ballen pattern.

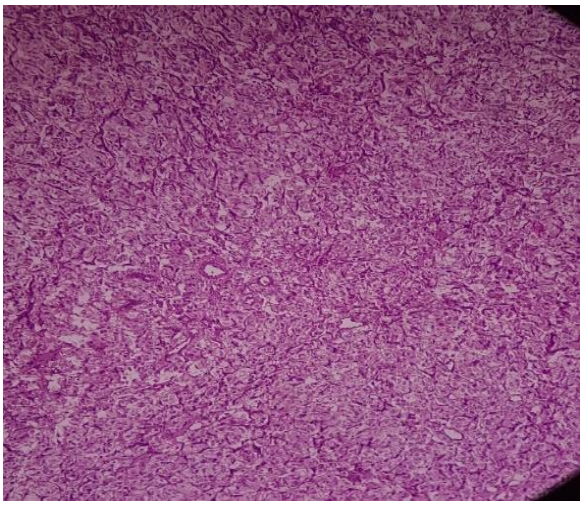

Figure-4: Right adrenal mass. Low power view (4 x) showing the nested pattern and packeting of cells.

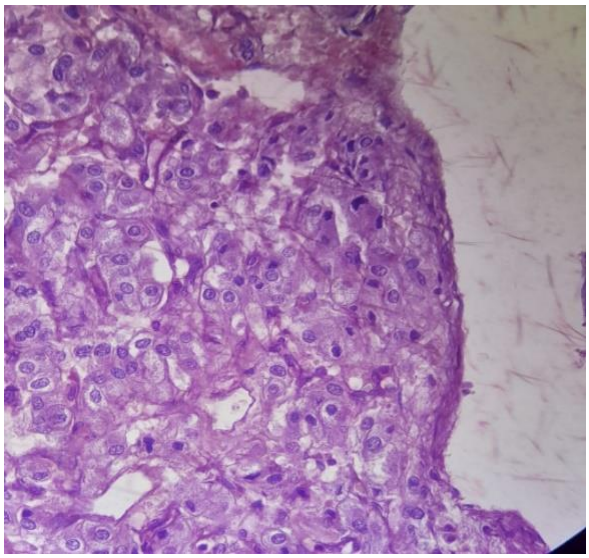

Figure-3: High power view (40x) with polygonal to round cells with abundant cytoplasm. An occasional mitotic figure is seen.

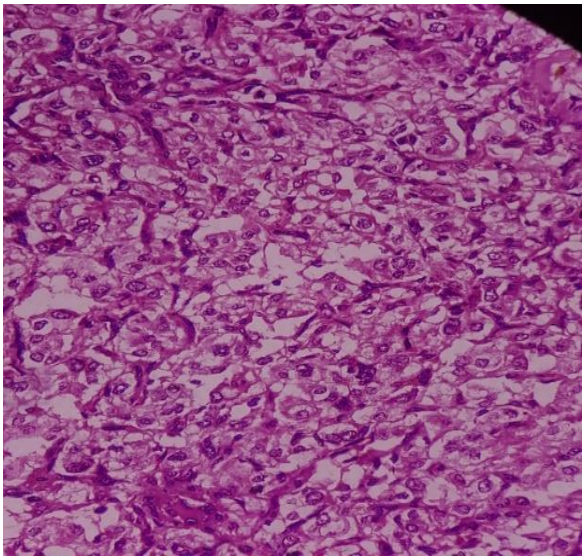

Figure-5: Right adrenal mass high power view (40x) showing polygonal cells arranged in packets with eosinophilic cytoplasm, minimal atypia. No mitotic figures.

After successful recovery from the left adrenalectomy, she underwent a right open adrenalectomy with a cut section showing a brownish encapsulated lesion having a central fibrotic bank. Histological examination showed tumor cells with abundant grey-blue granular cytoplasm and nuclei with granular chromatin, mitotic rate of less than 2 mitosis / 10HPF confirming the histopathological diagnosis of Left pheochromocytoma. (Figure 4, Figure 5)

In the post operative period, the patient had no major complications and was referred to a medical specialist on discharge for lifelong monitoring of Blood pressure and electrolyte balance who advised oral steroids. The six-month follow-up of patients with both us and the medical team was satisfactory.

DISCUSSION

With bilateral adrenal masses being rare, there have been very few documented cases of such nature.[1-3] Establishment of diagnosis can be difficult as tumors may be purely

incidental or may produce symptoms that overlap with a lot of other medical conditions which are generally ignored by patients including episodic headaches, sweating, tachycardia, flushing, paroxysms of hypertension.[3] Bilaterality poses an additional challenge of potential drastic metabolic and electrolyte disturbances that can be life threatening. Diagnostic workup may comprise Urinary catecholamine levels including metanephrines, normetanephrine and dopamine.

Further imaging workup can include Ultrasound Abdomen, CT Scan with contrast, MRI and MIBG scan where suited.[6-8] Bilateral resection of adrenal masses can be achieved in the same session by Laparoscopic approach or we may choose one by one approach through open adrenalectomy.

Histopathology of such rare cases requires a careful approach including a panel of immunostaining to confirm the diagnosis[9,10] as was done in this case. In such cases, a 2nd review from another pathologist in the same setup or outsourcing to obtain a second valid opinion should also be carried out for safe surgical practices and patient welfare, as was done in our case with both sources confirming the diagnosis of Pheochromocytomas.

There are very few cases being documented on bilateral pheochromocytomas and there is a need to gather more data to establish guidelines for diagnosis, histopathological reporting and treatment for such cases.

CONCLUSION

Bilateral Pheochromocytomas is a rare finding and poses a diagnostic and therapeutic challenge. The clinical presentation may be varied including presentation as incidentalomas. The use of a Contrast CT scan and Urinary catecholamines can narrow down the diagnosis with histopathology and immunocytochemistry staining being confirmatory and pathognomonic.

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AUTHOR'S CONTRIBUTION

NAG: Concept, Design and writing with data collection
SH: Data collection and critical review
UA: Data collection

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