A Young Man with a Rare Case of Adrenal Incidentaloma

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ABSTRACT

Adrenal incidentalomas (AIs) are adrenal masses discovered as incidental finding, often on imaging studies, unrelated to adrenal disorders. Sometimes, they are discovered as part of work-up for adrenal pathology. AIs are mostly non-functional (hormonally silent), but can also be functional (hormonally active). The differential diagnosis includes many primary, metastatic, benign and malignant conditions.

The current case is a young male, who went to a peripheral health facility with complaints of dysuria and burning micturition. His laboratory and radiological investigations suggested a urinary tract infection with an incidental finding of adrenal mass. After referral, his detailed work-up was done at Endocrine Clinic of Armed Forces Institute of Pathology (AFIP), Rawalpindi, which revealed history of episodic headaches and palpitations with paroxysmal spikes of high blood pressure up to 200/120 mmHg. Adrenalectomy was performed by laparoscopic surgery. Histopathological examination confirmed the diagnosis of pheochromocytoma with PASS score of 5/20. This is one of the rare cases of adrenal incidentaloma, often an autopsy finding. Pheochromocytoma needs to be investigated in all cases of AIs.

Key Words: Adrenal incidentaloma, Pheochromocytoma, Urinary tract infection.

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INTRODUCTION

Adrenal masses discovered incidentally are termed as adrenal incidentalomas (AIs). They are often discovered on imaging studies unrelated to adrenal pathology; and usually, the patient has no signs and symptoms of adrenal hormonal excess or obvious underlying malignancy. Less commonly, they are discovered as part of workup for adrenal disorders (e.g. Cushing’s, Pheochromocytoma etc.). The differential diagnosis of AIs includes many primary, metastatic, benign, and malignant entities arising from adrenal cortex or medulla. AIs are mostly non-functional (hormonally silent), while 15% cases are found to be functional, i.e., having active disease (e.g. pheochromocytoma, primary aldosteronism, Cushing’s, adrenal carcinomas, etc.), which require further evaluation and treatment to avoid medical complications. Work-up for AIs must be started to rule out any functional disorder and is invariably started with evaluation of adrenal profile. Serum cortisol, corticotrophin, plasma aldosterone, and renin levels should be assessed.

Pheochromocytoma being one of the most common among AIs must be evaluated by measuring 24 hours urinary vanillyl mandelic acid (VMA) and plasma metanephrine and non-metanephrine levels. As adrenal gland is the fourth common site for metastasis, work-up must include ruling out cancers of other organs as well as parathyroid hormone levels and calcitonin for pheochromocytoma associated malignancy (MEN2A).

CASE REPORT

A 25-year male presented with complaints of dysuria and burning micturition for 5 days at a peripheral hospital. Initial imaging by ultrasound kidney ureter bladder (USG KUB) revealed an adrenal mass. He was treated for urinary tract infection (UTI) and was referred to a tertiary care hospital for further work-up. Detailed evaluation was done when he presented to the Endocrine Clinic of Armed Forces Institute of Pathology (AFIP), Rawalpindi.

Detailed history revealed complaints of recurrent headache and palpitations for last 2 years. Headache was intermittent, moderate to severe in intensity, throbbing in character and was associated with photophobia, phono-phobia and dizziness. Palpitations were episodic, lasting 5-10 minutes, recurring 3-4 times daily with more frequency at night, associated with heat intolerance, sweating and feeling of chest tightness. There was no blurring of vision or any neurological, respiratory and gastrointestinal system related complaints. General physical examination revealed a young man of average built, conscious and well oriented, having tachycardia and high blood pressure occurring as paroxysmal attacks going up to 200/120 mmHg. Electrocardiogram showed sinus tachycardia with left ventricular hypertrophy.
Contrast enhanced computerised tomography (CECT) with adrenal protocol was carried out for confirming initial USG finding of adrenal mass, which revealed adrenal mass, measuring 4.4×5.4×6.5 cm (Figure 1). Further work-up for AI was done to rule out adrenal insufficiency, hypersecretion or any metastasis. Plasma aldosterone and morning cortisol levels along with low dose dexamethasone suppression test and plasma active renin concentration (ARC) were found normal, i.e., 385 pmol/L (normal: 140-830), 399 nmol/L (normal: 138-690), 49.5 nmol/l (normal: <50nmol/l) and 30.5 µU/mL (normal: 8-35), respectively. Tumor markers (AFP, CEA and CA 19-9) were evaluated for any underlying malignancy with adrenal metastasis, which were normal. Pheochromocytoma was suspected and 24 hours urinary vanillyl mandelic acid (VMA) levels were evaluated, which were surprisingly within normal physiological range i.e. 42 µmol/24hr (normal: 5-56). Free plasma metanephrine and nor-metanephrine levels were raised almost 10-folds, i.e., 5.3 nmol/L (normal: <0.5) and 8.3 nmol/L (normal: <0.9), respectively, leading to the diagnosis of pheochromocytoma.

Metabolism of dopamine involves dopamine-β-hydroxylase (DBH) and aromatic L-amino acid decarboxylase (AADC). In the absence of functional tumors, the activity of DBH and AADC is very low, but in pheochromocytoma, the activity of these enzymes is very high.

The malignant potential must be thoroughly evaluated. The treatment for a hormonally active (functional) adrenal tumor is surgery. The treatment for a hormonally silent adrenal incidentaloma is pheochromocytoma. Nonfunctional adrenal cortical adenoma can be diagnosed with a high degree of accuracy using imaging modalities. The adrenal gland is the fourth most common site of metastasis, and adrenal metastases may be found in as many as 25% of patients with known primary lesions. The treatment for a hormonally active (functional) adrenal tumor is surgery. The treatment for a malignancy depends on the cell type, spread, and location of the primary tumor. Nonfunctional adrenal cortical adenomas are not premalignant, and surgical excision is not indicated.

The patients with pheochromocytoma need to be treated with care as they present with vague symptoms and are easily missed. Multidisciplinary approach must be adopted which includes medical specialist, surgeon, anesthetist and pathologist. The malignant potential must be thoroughly evaluated. PASS score ≥4 suggests a malignant potential.

Pheochromocytoma should be considered in all AI cases because they are more common than previously thought, the diagnosis is often overlooked, and a failure to recognize them may lead to patient death.

CONFLICT OF INTEREST:
The authors declared no conflict of interest.

AUTHORS’ CONTRIBUTION:
UBK: Idea, data collection.
QA: Stastical analysis, data interpretation.
ZHH: Review and correction.
MA: Review and data interpretation.
SA: Discussion and preview.
SS: Literature review.
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