A rare silent pheochromocytoma: A diagnostic and management challenge

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Background:

Pheochromocytomas are extremely rare, usually non-malignant, tumors that develop in adrenal glands and result in excessive catecholamine production. Tumors usually present with paroxysmal or persistent hypertension, severe headache, palpitations, excessive sweating, tremors, and anxiety. Diagnosis is based on biochemical tests measuring catecholamines or their metabolites in urine or plasma and radiological imaging. The GAPP (Grading of Adrenal Pheochromocytoma and Paraganglioma) and PASS (Pheochromocytoma of the Adrenal Gland Scaled Score) grading systems help assess malignancy and recurrence.

Case presentation:

A 33-year-old man was admitted to an endocrinologist due to a left adrenal incidentaloma. Initially, the patient presented with abdominal pain in the left lower quadrant, and an abdominal ultrasound revealed an oval heterogeneous formation near the spleen's inferior pole, the etiology of which was unclear. Subsequent computed tomography (CT) of the upper abdomen confirmed an 11x8.8x11 cm formation in the left adrenal gland, primarily suggesting a pheochromocytoma. Notably, the patient exhibited no typical symptoms of pheochromocytoma. Hormonal and clinical examination excluded Cushing's syndrome. In addition, plasma normetanephrine and metanephrine levels were slightly elevated, but repeated biochemical tests showed results within the normal range. Adrenalectomy was performed, which confirmed a compound pheochromocytoma with a 5-10% ganglioneuromatous component. The GAPP (5/10) and PASS (7/20) scores indicated moderate differentiation and potential aggressiveness of the tumor, respectively. The patient was scheduled for a follow-up abdominal MRI and genetic testing.

Conclusion:

In the differential diagnosis of patients with silent pheochromocytoma, emphasis is placed on a comprehensive biochemical/radiologic workup to identify and treat this rare but potentially life-threatening condition accurately. The definitive diagnosis is confirmed through histopathologic findings following adrenalectomy. Additionally, calculating PASS and GAPP scores offers insights into the malignant potential and likelihood of recurrence. However, the follow-up of patients with silent pheochromocytoma poses significant challenges due to its unpredictable nature and asymptomatic presentation.

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