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Research Article

A Rare Case of Pheochromocytoma in The Elderly: The Importance of Additional Work-Up of Incidentalomas in Patients with Poorly Controlled Hypertension

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Introduction

Pheochromocytomas are rare catecholamine-secreting tumors primarily arising from the adrenal glands that are responsible for 0.05-0.2% cases of hypertension. The mean age at diagnosis is 42, and the actual incidence in very elderly patients is unclear [1,2]. Pheochromocytomas are often detected incidentally and pose a serious challenge to diagnose given that the symptoms are frequently paroxysmal and often attributed to other more common medical conditions such as essential hypertension and anxiety-related disorders. We present a rare case of symptomatic sporadic pheochromocytoma in a very elderly patient.

Case Report

An 86-year old female presented to the Emergency Department (ED) with palpitations, diaphoresis, hypertension, and intermittent chest pains. She also stated she had similar episodes during the past year. After initial workup, a CT of the chest and abdomen revealed a Thoracic Aortic Aneurysm (TAA), abdominal aortic aneurysm, and a 2.8cm enlarging unilateral adrenal mass. The patient's symptoms were attributed to the TAA by cardiology and appropriate surgical correction was carried out. Her recovery was complicated by an episode of severe hypertension (229/94 mmHg) and chest pain with EKG demonstrating T-wave abnormalities of the inferior leads as well as new second-degree type 1 atrioventricular block. The patient was subsequently diagnosed with NSTEMI and managed accordingly. The patient was lost to follow up soon after and returned 3 months later with focal neurological symptoms suggestive of Cerebrovascular Accident (CVA). During her hospital course, she had an episode of tachycardia with palpitations believed to be due to anxiety congruent with her past medical and social history. Therefore, anxiolytics were continued and metoprolol was added. Two months later, she presented to the ED with severe headache, chest pain, tremors, and a history of uncontrolled hypertensive episodes at her nursing facility. She was subsequently diagnosed with a subarachnoid hemorrhage along with poorly controlled hypertension. After witnessing several episodes of rapidly fluctuating blood pressures, serum metanephrines were ordered and found to be elevated. A subsequent workup including an MIBG and SPECT-CT scan confirmed the presence and location of a pheochromocytoma, which was later removed surgically. The diagnosis was confirmed by pathology via light microscopy and immunohistochemical staining, showing the mass to be chromogranin A and S-100 positive. The patient did well postoperatively and the hypertensive episodes, tachycardia, palpitations, and anxiety resolved (Figure 1,2).

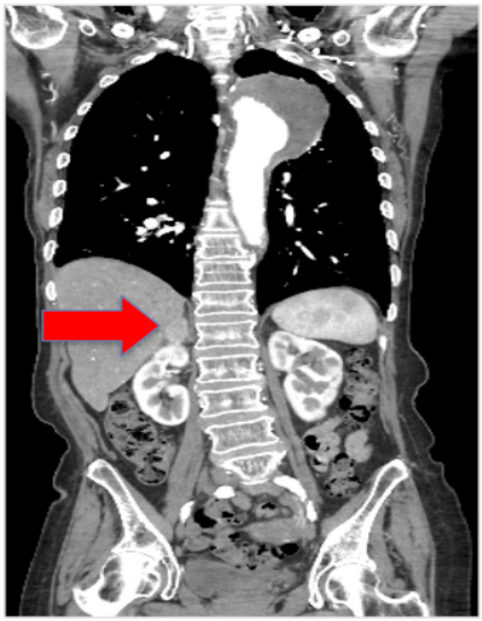

Figure 1: CT-chest/abdomen with contrast demonstrating 2.8 cm right adrenal mass.

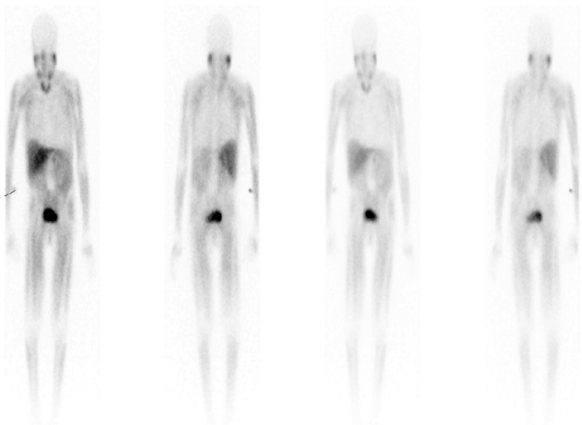

Figure 2: Whole body MIBG scan demonstrating both physiologic and abnormal uptake at 24 and 48 hours.

Discussion

The classic triad of pheochromocytoma involves episodic headache, sweating, and tachycardia. However, only about 50% of patients have paroxysmal hypertension at presentation [3]. Less common symptoms include orthostatic hypotension, blurred vision, weight loss, hyperglycemia, psychiatric disorders, and cardiomyopathy. In addition, the classic symptoms of pheochromocytoma discussed occur with less frequency when the tumor is associated with the familial multiple endocrine neoplasia type 2 as well as von Hippel-Lindau disease [4]. This highlights the necessity of performing genetic analysis in patients with pheochromocytoma, as many of the genetic syndromes associated with pheochromocytoma have multiple types of tumors in different areas of the body. In addition, genetic analysis would aid in determining the potential risks of other family members as well.

While pheochromocytoma is mostly diagnosed in middle-aged patients, some studies indicate that up to 50% of pheochromocytomas are diagnosed during autopsy [2]. This suggests that pheochromocytoma may be often undiagnosed in elderly patients due to atypical clinical presentation or because elderly patients may pose a more difficult challenge to diagnose, possibly because they often present with multiple comorbidities [5,6]. Furthermore, guidelines for the workup of incidentalomas suggest measurement of plasma-free metanephrines or urinary fractioned metanephrines, particularly with regards to tumors increasing in size and patients with symptoms suggestive of a catecholamine-secreting tumor [7]. In this case, the adrenal incidentaloma was determined to be enlarging based on prior abdominal CT scans in addition to the patient exhibiting episodic symptoms suggestive of catecholamine excess. These findings necessitate further workup involving plasma-free metanephrines or urinary fractioned metanephrines to assess for pheochromocytoma. In this particular case, the diagnosis of pheochromocytoma would have been eight months earlier-highlighting the importance of following guidelines regarding incidentalomas. This case also demonstrates how clinical decisions can be influenced by an anchoring effect, or the tendency to rely heavily on initial pieces of information when making clinical decisions [8]. The initial thought that her symptoms were cardiac/anxiety-related in nature were persistent throughout the nine months before diagnosis until she was reassessed by her healthcare team a for a secondary source of her poorly controlled hypertension.

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