Multisystem

Unsuspected pheochromocytoma incidentally found on chest CT

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ABSTRACT

A 51-year-old woman with history of migraine headaches and intermittent nausea, vomiting, palpitations, and diaphoresis presented to the emergency department with hypertensive emergency 1 month after starting a beta blocker for migraine prophylaxis. Contrast-enhanced computed tomography of the chest incidentally revealed a large abdominal mass in the area of the left adrenal gland. Iodine-123 metaiodobenzylguanidine scan imaging showed localized uptake into the left adrenal gland. Along with imaging results, laboratory testing confirmed the diagnosis of pheochromocytoma. The patient was treated with blood pressure control, specifically alpha blockade, and surgical excision of the mass. This case displays a typical clinical presentation of pheochromocytoma coupled with atypical radiographic size and appearance.

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Introduction

Pheochromocytomas are rare tumors derived from the chromaffin tissue in the adrenal medulla. Typically, pheochromocytomas are discovered at a size >3 cm, with an average of approximately 5 cm. The following is a case presentation of a large, 11.7 x 10.9 cm pheochromocytoma discovered incidentally during workup for hypertensive emergency. Given the tumor’s size, multiple imaging modalities were used to evaluate the extent of disease for preoperative staging and management. Ultimately, the patient underwent 14 days of phenoxybenzamine alpha blockade followed by resection of the mass.

Case report

A 51-year-old Caucasian female with no significant past medical history presented to an outside hospital complaining of severe headache and nausea as well as vision changes. Symptoms of intermittent nausea, vomiting, headache, diaphoresis, anxiety, and palpitations began 12 months earlier and had progressed over the 3 months prior to admission. For the month prior to admission, the patient received treatment from her primary care physician for migraines with propranolol and fioricet. On initial examination, she appeared anxious, complaining of a severe headache and color vision changes. She also had intractable nausea and vomiting, and was found to have a blood
pressure of 255/140 mm Hg in both arms with a heart rate of 150 beats per minute. She denied any fevers, chills, chest pain, or shortness of breath. Evaluation at an outside hospital included a computed tomography (CT) scan of the head, which was within normal limits, and a CT scan of the chest which was negative for aortic dissection but incidentally revealed a left upper quadrant abdominal mass measuring 10 cm × 12 cm. The mass was noted to compress the stomach and abutt the adrenals and spleen. On hospital day 2, she was stabilized on a sodium nitroprusside drip and transferred to our hospital for further management.

Upon admission to our institution, the patient endorsed mild nausea, vomiting, and central scotoma with color vision loss in the right eye. Initial vital signs showed a temperature of 99.0°F, a heart rate of 122 beats per minute, blood pressure of 180/140 mm Hg (equal in both arms), and 96% oxygen saturation on room air. On physical examination the patient appeared anxious but comfortable; she was alert and oriented to person, place, and time. Cardiovascular examination showed a normal S1, S2, no murmurs, gallops, rubs, heaves, thrills, or jugular venous distention. Peripheral pulses were palpable and there was no extremity edema. Abdominal examination was significant for mild, left upper quadrant tenderness, fullness to palpation, and a left-sided mid-abdominal bruit heard superior to the umbilicus. No distention or masses were noted on examination. Examination of the right eye revealed macular edema, flame hemorrhages, and a deficit in central visual field and a visual acuity of 20/70. The left eye had trace macular edema, fewer flame hemorrhages, and a visual acuity of 20/30.

Notable laboratory values on admission include potassium of 3.0 mEq/L (normal range 3.5-5.0), troponin I of 0.27 ng/mL (normal <0.05), lactate dehydrogenase 367 IU/L (normal 125-240), and brain natriuretic peptide 2654 pg/mL (normal <100). Imaging at the time of admission included a CT scan of the abdomen and pelvis, an upright chest x-ray, and a CT scan of the head. Significant positive and negative findings on the abdominal CT scan, as seen in Figures 1-6, include an 11.7 × 10.9 × 3.5-cm left upper quadrant abdominal mass with a feeding vessel arising directly off the abdominal aorta. The mass abutted abdominal viscera and was discerned anatomically to be originating from the adrenal gland. Laboratory evaluation of 24 hours of urine metanephrines and normetanephrines revealed elevations of 85,247 mcg (ref: 30-180) and 91,463 mcg (ref: 128-484), respectively. Subsequent fluorodeoxyglucose positron emission tomography-computed tomography (FDG-PET/CT) and iodine-123 metaiodobenzylguanidine (MIBG) scintigraphy, as seen in Figures 7-9, excluded malignancy and extra-adrenal foci. MIBG confirmed massive unilateral adrenal uptake compatible with pheochromocytoma. The patient was stabilized, completed a 14-day course of oral nonreversible alpha blockade, phenoxymenzamine, and underwent successful resection of her pheochromocytoma with resolution of all symptoms, Figures 10 and 11 show the gross specimen.

**Discussion**

Pheochromocytomas are rare tumors derived from the chromaffin tissue in the adrenal medulla which are capable of producing high levels of endogenous catecholamines. Although primarily benign and rare, with rates of occurrence between 2 and 8 per million people [1], pheochromocytomas are associated with high morbidity and mortality secondary to the effects of the catecholamines, including hypertension, cardiac arrhythmias, and end-organ damage.
to the physiological effects of catecholamines. Tumor cells release epinephrine and norepinephrine resulting in chronic refractory hypertension and eventually end organ damage. Acute release of hormones can precipitate hypertensive
emergency, stroke, myocardial infarction, and potentially death. Pheochromocytomas come to the attention of physicians clinically by refractory hypertension or episodic symptoms fitting the clinical profile. The use of selective beta blockers in patients with pheochromocytomas should be avoided due to the fact that it can cause unopposed alpha stimulation which can worsen the symptoms of a pheochromocytoma. It is believed that our patient’s presentation was exacerbated by the fact that she was started on a selective beta blocker in the time leading up to admission. The classic triad of signs and symptoms includes episodic headaches, tachycardia, and diaphoresis, with other symptoms including palpitations, tremor, dyspnea, and panic attack episodes from epinephrine secreting tumors [4].

Suspected pheochromocytomas should be evaluated first with laboratory testing such as 24 hours of urine collection for catecholamine by-products. Following biochemical confirmation of the tumor, radiological evaluation is necessary for localization. A CT of the abdomen is the first-line imaging modality with sensitivity of approximately 89% [5]. For a patient with no history of malignancy and an incidentaloma >4 cm, the American College of Radiology (ACR) appropriateness score of 8 out of 10 confirms abdominal CT scan with iodinated contrast as the first-line imaging test. Findings consistent with pheochromocytoma include a large heterogenous mass with areas of necrosis and cystic change. Given the high degree of vascularity, arterial and portal venous phases show increased...
enhancement that helps distinguish pheochromocytoma from adrenal adenoma. A value of approximately 110 Hounsfield units in the arterial phase is consistent with the diagnosis. Magnetic resonance imaging (MRI) of the abdomen has a sensitivity of 98% [5]. MRI of the abdomen with and without gadolinium contrast received an ACR appropriateness score of 8 and is recommended as an alternative to CT as part of preoperative staging.

Approximately 10% of pheochromocytomas are extra-adrenal [6], especially with large tumors (>10 cm) due to increased risk of malignancy or ectopic foci [7]. Iodine-123 MIBG scintigraphy may be useful for differentiation from paraganglioma or determining multifocality. MIBG is a compound similar to norepinephrine (with radiological properties) that undergoes uptake by catecholamine-producing tissue. MIBG scintigraphy is useful for detecting functional masses missed on CT-MRI or multiple smaller masses throughout the body. In patients with adrenal incidentalomas, with and without history of malignancy, MIBG scan received an ACR appropriateness score of 1 and 2 out of 10, respectively, with the

Fig. 9 – MIBG single photon emission computed tomography demonstrates avid unilateral radiotracer uptake in the large left adrenal mass, which is consistent with a pheochromocytoma. MIBG, metaiodobenzylguanidine.

Fig. 10 – Gross specimens show a large encapsulated and well-defined solid mass, with multiple engorged peripheral vessels coursing through the capsule.

Fig. 11 – Gross specimens show a large encapsulated and well-defined solid mass, with multiple engorged peripheral vessels coursing through the capsule.
recommendation to perform the scan only for suspicion for pheochromocytoma. It carries a high degree of radiation exposure. For our patient, it was useful to perform an MIBG scan to ensure that the patient did not have any evidence of extra-adrenal disease. Once confirmed, the patient was able to undergo definitive treatment with surgical excision of the pheochromocytoma.

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