CLINICAL VIGNETTE

Pheochromocytoma

Matteo Dinolfo, M.D.

Case Presentation

A 58-year-old male with history of COPD presented with two weeks coughing and increased blood pressure. The patient denied fever, but his wife reported increased sweating for the past month. He denied headache, tachycardia, diarrhea or anxiety.

Additional past history includes current smoking, gastroesophageal reflex and Vitamin D deficiency. Current medications were omeprazole and Vitamin D3.

On physical examination, blood pressure was 174/96, pulse 78 with respirations of 14/min. Exam was remarkable for rhonchi in the left upper lung field. Cardiac and abdominal exams were normal without murmurs or hepatosplenomegaly.

Initial diagnostic workup included normal CBC and chemistries. Chest x-ray showed hyperinflation consistent with COPD with a left upper lobe density, scar vs. mass.

Follow-up chest CT showed COPD, bronchiolitis with incidental finding of a 2x2 cm left adrenal mass. Twenty-four hour fractionated metanephrines were 2000 mcg/24 hours, with normal plasma free metanephrines.

Surgical and Endocrine consultations were obtained with presumptive diagnosis of pheochromocytoma.

Preoperatively the patient was treated with alpha adrenergic blockade with Phenoxybenzamine 20 mg BID.

He underwent successful laparoscopic resection of the adrenal tumor. Pathology confirmed pheochromocytoma. His postoperative course was uncomplicated without hypertensive crisis and his blood pressure normalized off medications.

Discussion

Pheochromocytoma are catecholamine secreting tumors of the chromaffin cells of the adrenal medulla. They may also occur extrarenal. Malignant and benign tumors appear the same pathologically. Pheochromocytomas are rare tumors occurring in about 0.2% of patients with hypertension with annual incidence of 0.8 per hundred thousand patient years. They can occur at any age but most often in the fourth or fifth decade with equal incidence in males and females.

The classical triad of symptoms consists of episodes of headaches, sweating and tachycardia. Fifty percent have paroxysmal hypertension, the remainder with persistent hypertension. The majority of patients do not have classical symptoms. Less common symptoms include orthostatic hypotension, visual blurring, weight-loss, polyuria, polydipsia and papilledema.[1,2]

Approximately 10% of tumors are malignant although they are histological and biochemically identical. The diagnosis is confirmed by measurement of fractionated urinary and plasma metanephrines.

Tricyclic antidepressants may interfere with the interpretation of urinary catecholamines.[3]

Treatment

Laparoscopic resection of the adrenal mass with alpha adrenergic blockade and volume expansion is the treatment of choice. Adrenal resection is a high risk surgical procedure and agents that can precipitate a hypertensive crisis should be avoided. These include glucagon, histamine and metoclopramide.

Alpha adrenergic blockade should precede surgery by 10 to 14 days. The initial dose of phenoxybenzamine is 10mg twice daily with increase to 10-20 mg in divided doses every 2 to 3 days. Typical final dose is 20-100 mg daily. Other selective alpha-adrenergic blocking agents such as prazosin, terazosin or doxazosin are also utilized in many centers particularly in metastatic pheochromocytoma.

Beta-adrenergic blockers should never be started prior to alpha-adrenergic blockers because the blockade of vasodilatory peripheral beta adrenergic receptors with unopposed alpha-adrenergic receptor stimulation can lead to a further increase in blood pressure.[4]

Conclusion

This hypertensive patient was discovered to have an adrenal mass as an incidental finding on CT imaging for an unrelated

problem. Prompt evaluation with urinary metanephrines and appropriate surgical and endocrine management resulted in an excellent outcome.

REFERENCES

1. **Young, William F. Jr**. Adrenal causes of hypertension: pheochromocytoma and primary aldosteronism. *Review of Endocrine and Metabolic Disorders*. 1007; 8:301.
2. **Stein PP, Black HR**. A simplified diagnostic approach to pheochromocytoma. A review of the literature and report of one institution's experience. *Medicine*; Medicine;1991-70:40.
3. **Young WF Jr**. Clinical Practice. The incidentally discovered adrenal mass. *N England Journal of Medicine*. 2007:356:601.
4. **Young William M.D.; Norman M Kaplan, M.D.Electron Kebebew,M.D.**; Treatment of pheochromocytoma in adults. Official reprint of up to Date 2014.

Submitted on September 5, 2014