AN INTERESTING CASE OF HYPERTENSION

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Abstract:
Hypertension is a common disease which we encounter day by day. Most of the time the cause is not known and are labeled as essential or primary hypertension. A small percentage of patients (2-10) have a secondary cause. Here is a 42 year old male presenting with complaints of chest pain for 3 days with history of giddiness, palpitation, excessive sweating, headache and blurring of vision. He also had fear during these symptoms. On examination he had tachycardia and hypertension. His systemic examination was normal. Complete hemogram, RFT, LFT were within normal limits. USG abdomen showed right adrenal mass, CECT abdomen also showed right adrenal mass. 24 hours urine VMA were raised, MIBG scan findings are compatible with pheochromocytoma. Hypertension and tachycardia with adrenal mass helped us to come to a conclusion of pheochromocytoma.

Keyword
Hypertension, Pheochromocytoma, Paraganglioma

CASE SUMMARY

46 year old male admitted with complaints of chest pain for past 3 days. He also had history of giddiness, palpitation, excessive sweating, headache and blurring of vision. History of fear present during these symptoms. He is not a known diabetic, hypertensive, tuberculosis, bronchial asthma; CAD. He is a known alcoholic for past 15 years.

On examination, pulse 130/min, BP 190/120mmHg, Systemic examination of CVS, RS, Abdomen, CNS were normal, fundus examination showed grade 2 hypertensive retinopathy. With these features we arrived at a provisional diagnosis of systemic hypertension probably due to secondary cause. Investigations revealed CBC, RFT, LFT were within normal limits, ECG showed sinus tachycardia, chest X-ray normal, echocardiography normal. USG abdomen showed right adrenal mass. He was treated with tablet amlodipine 10mg and symptomatically. In view of suspected secondary hypertension we proceeded with renal Doppler, which was normal. Serum cortisol and ACTH were normal, 24 hours urine vanillylmandelic acid (VMA) was increased, 32.2mg/24hours (0-13.6mg/24 hours). CECT
abdomen showed heterogeneously enhancing lesion of right adrenal gland.

We further proceeded with I\textsuperscript{131} MIBG and the scan findings are compatible with pheochromocytoma of right adrenal gland.

The final diagnosis was pheochromocytoma causing secondary hypertension.

Patient was treated with Cap. Phenoxybenzamine 10mg TDS, Tab. Propranolol 40mg BD and liberal salt intake. With this treatment patient symptomatically improved and his blood pressure came to 130/80mmHg and pulse 78/min. In view of possibility of MEN 2 syndromes MRI brain was taken which was normal. Patient underwent adrenalectomy successfully.

DISCUSSION:
Pheochromocytomas and paragangliomas are catecholamine-producing tumours derived from the sympathetic or parasympathetic nervous system. These tumours may arise sporadically or be inherited as features of multiple endocrine neoplasia type 2 or several other pheochromocytoma-associated syndromes. The diagnosis of pheochromocytomas provides a potentially correctable cause of hypertension, and their removal can prevent hypertensive crises that can be lethal\textsuperscript{1}. The clinical presentation is variable, ranging from an adrenal incidentaloma to a patient in hypertensive crisis with associated cerebrovascular or
cardiac complications. Pheochromocytoma is estimated to occur in 2–8 of 1 million persons per year, and about 0.1% of hypertensive patients harbour a pheochromocytoma. Autopsy series reveal prevalence of 0.2%. The mean age at diagnosis is about 40 years, although the tumours can occur from early childhood until late in life. The "rule of tens" for pheochromocytomas states that about 10% are bilateral, 10% are extra adrenal, and 10% are malignant. However, these percentages are higher in the inherited syndromes. Pheochromocytomas and paragangliomas are well-vascularized tumours that arise from cells derived from the sympathetic (e.g., adrenal medulla) or parasympathetic (e.g., carotid body, glomus vagale) Para ganglia. The name pheochromocytoma reflects the black-colored staining caused by chromaffin oxidation of catecholamines. Although a variety of terms have been used to describe these tumours, most clinicians use the term pheochromocytoma to describe symptomatic catecholamine-producing tumours, including those in extra adrenal retroperitoneal, pelvic, and thoracic sites. The term paraganglioma is used to describe catecholamine-producing tumours in the head and neck. These tumours may secrete little or no catecholamines. The etiology of sporadic pheochromocytomas and paragangliomas is unknown. However, about 25% of patients have an inherited condition, including germ-line mutations in the RET, VHL, NF1, SDHB, SDHC, SDHD, or SDHAF2 genes. The clinical presentation is so variable that pheochromocytoma has been termed "the great masquerader". Among the presenting symptoms, episodes of palpitations, headaches, and profuse sweating are typical and constitute a classic triad. The presence of all three symptoms in association with hypertension makes pheochromocytoma a likely diagnosis. However, a pheochromocytoma can be asymptomatic for years, and some tumours grow to a considerable size before patients note symptoms. The dominant sign is hypertension. Classically, patients have episodic hypertension, but sustained hypertension is also common. Catecholamine crises can lead to heart failure, pulmonary edema, arrhythmias, and intracranial haemorrhage. During episodes of hormone release, which can occur at very divergent intervals, patients are anxious and pale, and they experience tachycardia and palpitations. These paroxysms generally last less than an hour and may be precipitated by surgery, positional changes, exercise, pregnancy, urination (particularly bladder pheochromocytomas), and various medications (e.g., tricyclic antidepressants, opiates, metoclopramide). Pheochromocytomas and paragangliomas synthesize and store catecholamines, which include norepinephrine (noradrenaline), epinephrine (adrenaline), and dopamine. Elevated plasma and urinary levels of catecholamines and the methylated metabolites, metanephrines, are the cornerstone for the diagnosis. The hormonal activity of tumors fluctuates, resulting in considerable variation in serial catecholamine measurements. Thus, there is some value in obtaining tests during or soon after a symptomatic crisis. However, most tumors continuously leak O-methylated metabolites, which are detected by measurements of metanephrines. Catecholamines and metanephrines can be measured by using different methods (e.g., high-performance liquid chromatography, enzyme-linked immunosorbent assay, and liquid chromatography/mass spectrometry). In a clinical context suspicious for pheochromocytoma, when values are increased three times.
the upper limit of normal, a pheochromocytoma is highly likely regardless of the assay used. However, the sensitivity and specificity of available biochemical tests vary greatly, and these differences are important in assessing patients with borderline elevations of different compounds. Urinary tests for vanillylmandelic acid (VMA), metanephrines (total or fractionated), and catecholamines are widely available and are used commonly for initial testing. Among these tests, the fractionated metanephrines and catecholamines are the most sensitive. Plasma tests are more convenient and include measurements of catecholamines and metanephrines. A variety of methods have been used to localize pheochromocytomas and paragangliomas. CT and MRI are similar in sensitivity. CT should be performed with contrast. T2-weighted MRI with gadolinium contrast is optimal for detecting pheochromocytomas and is somewhat better than CT for imaging extra-adrenal pheochromocytomas and paragangliomas. About 5% of adrenal incidentalomas, which usually are detected by CT or MRI, prove to be pheochromocytomas after endocrinologic evaluation.

Tumors also can be localized by using radioactive tracers, including 131I- or 123I-metiodobenzylguanidine (MIBG), 111In-somatostatin analogues, or 18F-dopa (or dopamine) positron emission tomography (PET). Because these agents exhibit selective uptake in paragangliomas, nuclear imaging is particularly useful in the hereditary syndromes. Complete tumour removal is the ultimate therapeutic goal. Preoperative patient preparation is essential for safe surgery. Adrenergic blockers (phenoxybenzamine) should be initiated at relatively low doses (e.g., 5–10 mg orally three times per day) and increased as tolerated every few days. Because patients are volume-constricted, liberal salt intake and hydration are necessary to avoid orthostasis. Adequate alpha blockade generally requires 7 days, with a typical final dose of 20–30 mg phenoxybenzamine three times per day.

Oral prazosin or intravenous phentolamine can be used to manage paroxysms while awaiting adequate alpha blockade. Before surgery, blood pressure should be consistently below 160/90 mmHg, with moderate orthostasis. Beta blockers (e.g., 10 mg propranolol three to four times per day) can be added after starting alpha blockers and increased as needed if tachycardia persists. Other antihypertensives, such as calcium channel blockers or angiotensin-converting enzyme inhibitors, have been used when blood pressure is difficult to control with phenoxybenzamine alone. Blood pressure can be labile during surgery, particularly at the onset of intubation or when the tumour is manipulated. Nitroprusside infusion is useful for intraoperative hypertensive crises, and hypotension usually responds to volume infusion.

CONCLUSION: Pheochromocytoma is one of the cause for secondary hypertension. History, clinical examination and relevant investigations are necessary to diagnose secondary hypertension. Always look for treatable cause for hypertension in all cases of hypertension.

REFERENCES:


