



The Great Masquerader

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Abstract

Pheochromocytoma is a rare neuroendocrine tumour composed of chromatin cells which secrete catecholamines. These tumours present with Menrad's clinical triad (headache, sweating and palpitations). Mainly located in the adrenal gland, they are more frequent between the 3rd and 5th decades of life. We present a case where a 33 year old female presents with an episode of seizure along with hypokalemia, paroxysmal hypertension with an adrenal mass on the right side. The patient was treated with alpha and beta blockers with surgical resection of the tumour and subsequent resolution of hypertension.

Case Report

A 33 year old female who is a housewife, presented to the ER around 5:30AM with 1 episode of generalised tonic clonic seizure which occurred at around 4am. Patient was brought to the ER in a post-ictal state. On examination, patient was drowsy and arousable. Her pulse rate was 124/min, regular in rhythm and normal in character. Her blood pressure was 110/70 mmHg. Her cardiovascular system and respiratory system were unremarkable. Her abdomen was soft and bowel sounds were heard. She was moving all 4 limbs and obeying simple commands. Initial Arterial blood gas done showed metabolic alkalosis

with hypokalaemia - pH 7.60, pCo₂ 40.8 mmHg, pO₂ 130.6 mmHg, HCO₃ - 40.8 mEq/L, K- 2 mEq/L, lactates 1.5 mmol/L. Capillary blood glucose on arrival was 146 mg/dL. ECG showed sinus tachycardia with evidence of left ventricular hypertrophy. An urgent CT brain was done which was normal. Patient was given loading dose of Fosphenytoin and initiated on parental potassium correction. Patient was then shifted to the intensive care unit for further management. Ultrasound abdomen was done showed bulky kidneys, CMD was maintained. Her hemoglobin was 9.4 g/dL, total white cell count was 23,700 cumm - polymorph predominant, platelets were 4.39 lakhs.

Her blood urea nitrogen was 15 mg/dL, creatinine was 2.1 mg/dL, sodium 131 mmol/lit, potassium 2.8 mmol/L, chloride 76 mmol/lit, bicarbonate 37 mmol/lit. Her liver function tests were within normal limits. Her urine routine had 6-8 pus cells with trace proteinuria. Her urine and blood cultures were sent.

Patient is a known case of diabetes mellitus, systemic hypertension diagnosed 2 months ago not on regular treatment. She initially had palpitations and grade 1-2 NYHA dyspnea on exertion for which she consulted a doctor after which she was initiated on antihypertensives but she was irregular with her medications.

She had been admitted 20 days ago in an outside hospital for accelerated hypertension, treated for the same and discharged after 5 days.

Patient then developed a urinary tract infection and was admitted in an outside hospital and was treated with intravenous antibiotics and discharged after 5 days.

With this history, a provisional diagnosis of acute pyelonephritis, acute kidney injury, new onset seizure with a background history of type 2 diabetes mellitus and systemic hypertension was made. As the patient had hypokalaemia, systemic hypertension and metabolic alkalosis, a possibility of primary hyperaldosteronism and pheochromocytoma were considered and hence worked up. Patient underwent CT abdomen which showed - a well defined lesion of size 33 x 33 x 42mm in the right suprarenal region, bilateral bulky kidneys without obvious fat stranding. Her sensorium had improved to a GCS of 15/15 and was symptomatically better.

A 24 hour urinary metanephrine and normetanephrine was sent. Potassium and magnesium supplementation was continued till hypokalaemia got corrected after which a plasma renin activity and plasma aldosterone ratio was sent.

Patient suddenly developed chest pain the next morning with a blood pressure of 250/120 mmHg. An ECG was done which showed new onset ST sagging in inferior leads. An urgent cardiology

opinion was obtained and a 2D Echocardiography was done. 2D echocardiography done showed a dilated left ventricle, concentric left ventricular hypertrophy, structurally normal valves, entire septum and anterior wall hypokinetic, moderate left ventricular systolic dysfunction (ejection fraction - 38%), IVC - 1cm, collapsing. Dissection of flap observed in descending aorta (double lumen). Trop I was 0.37. Patient was advised a CT aortogram. After nephrology clearance, patient underwent a CT aortogram. Patient was initiated on labetalol infusion.

CT aortogram showed no evidence of aortic dissection. However it showed a well defined heterogeneously enhancing lesion measuring 33 x 33 x 37 mm was noted arising from the right suprarenal gland with extensions as follows:

Superiorly and laterally abutting segment V of the liver

Inferiorly it is seen to efface the upper pole of the right kidney

Medially it is seen to cause extraneous compression of the infra hepatic IVC,

Fat plane is preserved within the surrounding structures.



Suggestive of right pheochromocytoma.

A diagnosis of probable stress cardiomyopathy was made and patient was initiated on supportive measures.

Her renal function tests were monitored daily and was found to be improving. Her total counts were

improving. She had a good urine output. Her urine and blood cultures showed no growth.

Her blood pressure was stable with prazosin and labetalol infusion as required. Patient was monitored in the intensive care unit. Her left ventricular function was monitored and found to be improving.

Her 24 hour urine metanephrine level was 1.69 mg/day (<1), normetanephrine was 2902 mcg/24 hours (<600), Plasma renin activity was 73.61 IU/ml (4.4 - 46.1), Plasma aldosterone was 13.50ng/dL (2.52 - 392.2), plasma metanephrine was 68 (<65) pg/ml, serum cortisol was 25.3mcg/dL.

A diagnosis of pheochromocytoma was confirmed. Patient underwent right adrenalectomy.

Histopathological examination of the adrenal gland showed sections of a well circumscribed capsulated tumour. Tumour cells were arranged predominantly in Zell ballen pattern with occasional trabecular and cords of cells. The individual cells are polygonal with abundant granular eosinophilic cytoplasm with a centrally located round and regular nucleus and with stippled chromatin. There is a rich vascular network surrounding the tumour nests. No evidence of capsular or vascular invasion. No evidence of increased mitosis, atypia or necrosis in the section examined.

Pheochromocytoma of the adrenal gland scaled score (PASS) was 2.

Hence a final diagnosis of pheochromocytoma was confirmed with PASS score 2.

Patient was then shifted to the ward for observation. Patient required anti hypertensives for 4 weeks after which it was stopped. Her repeat 2D echocardiography showed an ejection fraction of 55%. Patient has come for review and is currently doing well not requiring any medications.

Discussion

Pheochromocytoma is a rare neuroendocrine tumour composed of chromaffin cells which secretes catecholamines. Especially with young sub-

ject, this diagnosis is most often sought for in the context of a paroxysmal or permanent arterial hypertension, particularly when it is associated with Menard's clinical triad (headache, sweating and palpitations) ⁽¹⁾.

Nearly 80–85% of pheochromocytoma arise from the adrenal medulla, whereas about 15–20% are from the extra-adrenal chromaffin tissue ⁽²⁾.

Pheochromocytoma is sometimes called the "10% tumor" because approximately 10% are bilateral (i.e. in both adrenal glands), 10% are found in children, 10% are genetic (i.e. inherited), 10% are cancer, and 10% are found outside the adrenal gland (i.e. paraganglioma). Extra-adrenal pheochromocytomas are more likely to be malignant than adrenal pheochromocytomas. Malignant pheochromocytomas may spread (metastasize) to various areas of the body including the lymph nodes, liver, lungs, and bones.

Symptoms associated with pheochromocytomas occur because of the release of catecholamines (e.g., norepinephrine and epinephrine). Systemic hypertension is the most common finding associated with pheochromocytomas. High blood pressure may be paroxysmal - the frequency of these episodes vary anywhere from several times a day to a couple of times a month.

Additional symptoms that occur less frequently may include pain in the chest pain, abdominal pain, nausea, vomiting, diarrhea, constipation, pale skin (pallor), weakness, and weight loss. Attacks of anxiety or apprehension may also occur. Some individuals experience an extreme drop in blood pressure upon standing suddenly, sometimes resulting in dizziness (orthostatic hypotension). In some cases, individuals with a pheochromocytoma may be asymptomatic.

If left untreated, pheochromocytomas may progress to cause serious, life-threatening complications including cardiomyopathy, myocarditis, cerebral haemorrhage or acute pulmonary edema. Some patients may developed acute coronary syndrome.

As this patient had morphological abnormalities along with hypokalaemia and systemic hypertension, three diagnoses must be mentioned: a primary hyperaldosteronism, a subclinical cushing and finally a pheochromocytoma. The aldosterone/PRA ratio has the best diagnostic performance to screen for primary aldosteronism. It's considered as the pivotal test for the etiological diagnosis of hypertension with hypokalemia⁽³⁾, and the test of urinary metanephrines and normetanephrines will lead to diagnosis a pheochromocytoma⁽⁴⁾ as was the case of our patient.

CAUSE OF HYPERTENSION WITH HYPOKALEMIA
Essential hypertension with diuretic use
Primary aldosteronism
Pheochromocytoma
Cushing's disease
Renal Vascular disease
Malignant hypertension

The curative treatment of pheochromocytoma is based on surgical excision of the tumour. The frequency of recurrences, as well as the risk of delayed metastasis, can appear after some decades following the initial surgery. That's why the long-term monitoring of patients is required.

References

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