

CONCLUSION

In a highly suspicious case of primary aldosteronism, a repeat screening test is warranted to prevent missing the diagnosis. Performing screening tests for PA in ESRD can be attempted but expect complexity in interpretation. Spironolactone can be given with caution in ESRD patients with PA.

EP_A005

DIABETIC KETOACIDOSIS (DKA) AS A RARE PRESENTATION OF PHEOCHROMOCYTOMA

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INTRODUCTION/BACKGROUND

Pheochromocytoma commonly presents with hypertension. Diabetes mellitus is one of the extremely rare metabolic complications of pheochromocytoma and is seen in a third of patients with pheochromocytoma. We present a patient with pheochromocytoma whose initial presentation was DKA.

CASE

A 39-year-old Indian female presented with abdominal pain and fever and was diagnosed with DKA. Her weight was 40kg with BMI of 17kg/m². Her blood pressure was 90/60 mmHg. Underlying sepsis was suspected in the presence of leucocytosis (WBC 22x10⁹/L). Abdominal ultrasound showed a solitary liver lesion at segment V/V1. The CECT revealed a well-defined capsulated right suprarenal mass measuring 5.4 x 6.2 x 7.8 cm. Urine epinephrine level was elevated, 117.8 ug/day (0.5-2 ug/day), while both metanephrine and dopamine levels were normal. A diagnosis of right adrenal pheochromocytoma was made.

She was lost to follow-up but continued her diabetes management in the primary care clinic and remained on basal-bolus insulin. Four years later, she presented again with right-sided abdominal discomfort, with episodes of headache, palpitations and sweating. She also developed hypertensive crisis during this admission. CECT showed a large heterogeneously enhancing right suprarenal mass measuring 7.7 x 8.1 x 10.4 cm with mass effect to the inferior border of the liver and displacement of the right kidney with no evidence of distant metastasis. Urine metanephrine level was elevated at 82.2umol/day (0-1.62 umol/day) while urine normetanephrine level was 10.0 umol/day (0-2.13 umol/day)

She underwent right adrenalectomy. Post-operatively, she was euglycemic and normotensive and was discharged without any antihypertensives and insulin.

CONCLUSION

Pheochromocytoma rarely presents with DKA. The presence of DM in a young, lean patient might be the clue. Hypertension might not be present in the beginning as in this patient. Close glucose monitoring intra- and post-operatively is important as hypoglycaemia may occur after tumour resection.

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MAX AND HIS FURY SPELLS: A CASE OF BILATERAL PHEOCHROMOCYTOMA WITH MAX-ASSOCIATED PATHOGENIC GENE MUTATION

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INTRODUCTION/BACKGROUND

Understanding of the genetic pathophysiology of pheochromocytomas and paragangliomas (PPGLs) syndrome has advanced significantly over the past two decades. PPGLs entail three specific disease clusters based on their underlying genetic alterations. Pathogenic variants affecting the Myelocytomatosis-Associated factor X (MAX) gene predispose to PPGLs occurring at a younger age. More than half develop bilateral pheochromocytomas with metastatic disease seen in 20 percent of patients.

CASE

This is the first case report in Malaysia describing a young male with bilateral pheochromocytoma secondary to a novel pathogenic variant identified in the MAX gene.

A 28-year-old male was found to be hypertensive during a dental procedure. Four months later, he was hospitalized due to palpitations and treated for rhabdomyolysis with non-ST-elevation myocardial infarction. Echocardiography did not show cardiomyopathy and coronary angiography was normal. Endocrine evaluation showed an elevated 24-hour urine metanephrine level of 38.8 micromol/day (24 times ULN), urine normetanephrine level of 30.8 micromol/day (14.5 times ULN), and urine methoxytyramine level of 6.5 micromol/day (3.6 times ULN). Adrenal CT revealed bilateral lipid-poor adrenal masses (Left: 7.1 x 7.5 x 7.4 cm; and right: 2.0 x 1.1 x 1.8 cm). There was no family history of multiple endocrine neoplasia or Von-Hippel