## EP_A003

### 'AN IMPENDING DOOM:' RARE CASE OF RUPTURED PHEOCHROMOCYTOMA PRESENTING AS PHEOCHROMOCYTOMA CRISIS

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**INTRODUCTION/BACKGROUND**
Spontaneously ruptured pheochromocytoma presenting as pheochromocytoma crisis is a rare complication and is associated with high morbidity and mortality.

**CASE**
We present a case of a 46-year-old female with left pheochromocytoma, diagnosed following a total abdominal hysterectomy for uterine leiomyoma. 24-hour urine catecholamines were elevated- norepinephrine: 2.6 x ULN [1241 nmol/L], epinephrine: 12.8 x ULN [1468.3 nmol/L], dopamine <424 nmol/L. Plasma normetanephrine and metanephrines were also elevated. CT scan showed a large adrenal mass measuring 4.98 x 5.4 x 5.5 cm. Medications were oral Prazosin 3 mg five times daily and Labetalol 100 mg twice daily. She presented four weeks later with acute onset of abdominal pain, persistent vomiting, chest discomfort, vasovagal syncope, headache and sweating. She was restless, pale and hypotensive with a BP of 80/40 mmHg and HR 104 beats per minute. Abdominal examination revealed generalised tenderness, guarding and a palpable mass over the left lower quadrant. Blood pressure increased subsequently ranging from systolic 150 to 200 mmHg and diastolic 90 to 100 mmHg. ECG showed widespread deep T inversion suggestive of Wellen syndrome with raised Troponin I (1950 ng/mL). Abdominal CT revealed a ruptured left pheochromocytoma measuring 5.0 x 6.6 x 7.1 cm with a large intra-abdominal hematoma. She was started on Prazosin 2 mg three times daily and Labetalol 100 mg three times daily for blood pressure control. She was given an insulin infusion for hyperglycemia. Packed cells were also transfused. She underwent laparotomy and adrenalectomy following adequate alpha- and beta-blockade. The postoperative course was uneventful. HPE of the left adrenal mass confirmed pheochromocytoma.

**CONCLUSION**
Pheochromocytoma crisis resulting from a large release of catecholamines from a ruptured pheochromocytoma is associated with high mortality. Prompt resuscitation and blood pressure control are the mainstays of treatment prior to surgical intervention.

## EP_A004

### UNMASKING PRIMARY ALDOSTERONISM IN A PATIENT WITH END STAGE RENAL DISEASE: A CASE REPORT

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**INTRODUCTION/BACKGROUND**
Managing hypertension in a patient with end-stage renal disease (ESRD) requires a combination of antihypertensive medications and volume control. It is common to encounter refractory hypertension in ESRD. Investigation of primary aldosteronism (PA) as a cause of refractory hypertension in ESRD is potentially difficult. These patients are on multiple antihypertensive medications that cannot be discontinued, thus complicating the interpretation of aldosterone-renin ratio.

**CASE**
We present a patient with ESRD on peritoneal dialysis with refractory hypertension and hypokalaemia investigated for primary aldosteronism.

The patient is a 72-year-old male, known hypertensive for 40 years, with poorly controlled blood pressure for the past 20 years. The presence of refractory hypertension and hypokalaemia prompted an investigation for primary aldosteronism ten years ago, where the patient tested negative. Over the next ten years, his eGFR deteriorated, and he was initiated on peritoneal dialysis a year ago. Despite peritoneal dialysis, his BP remained poorly controlled while on six antihypertensive medications, including furosemide and spironolactone.

Despite being on six confounding antihypertensive medications, his plasma aldosterone was not suppressed and instead, elevated at 1229 pmol/L with a normal direct renin level (9 mU/L). Adrenal CT revealed bilateral adrenal adenomas. Further assessment with adrenal vein sampling was done. Surgery was explored, but the patient was not keen. Spironolactone dose was optimized which led to improvement of blood pressure control and reduction of other antihypertensive medication doses without occurrence of hyperkalemia.

**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/m$^2$. Her blood pressure was 90/60 mmHg. Underlying sepsis was suspected in the presence of leucocytosis (WBC 22x10$^9$/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.

## EP_A006

### 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