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

Lindau syndrome. Thyroid ultrasonography and retinal examination were normal. Our clinical dilemma was whether both adrenal lesions were pheochromocytomas. Gallium-68 PET/CT showed significant uptake in the left adrenal mass, indeterminate on the right. A genetic study identified a pathogenic variant c.234_235dup in exon 4 of the MAX gene. Henceforth, bilateral pheochromocytoma was highly considered. Because of the metastatic potential of the disease, he underwent bilateral adrenalectomy.

CONCLUSION

Each PPGLs cluster has a unique clinical, biochemical and imaging phenotype which can help clinicians deliver a personalized treatment strategy for patients with PPGLs. Precision medicine approach to PPGLs should be more widely available and become the standard of care in our nation.

EP_A007

LATERAL APPROACH RETROPERITONEOSCOPIC ADRENALECTOMY: A SINGLE-CENTRE MALAYSIAN EXPERIENCE

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INTRODUCTION

Laparoscopic or retroperitoneoscopic adrenalectomy is now the preferred approach when adrenalectomy is indicated. We report the first series of Lateral Approach Retroperitoneoscopic Adrenalectomy.

METHODOLOGY

Between 2013 and 2023, a total of 46 patients (28 males,18 females; mean age 48.4 years) were referred for minimally invasive adrenalectomy. The surgeon, a urologist, was trained in minimally invasive surgery and is a proponent of the lateral approach retroperitoneoscopic technique. Twenty-six cases involved the left side and 20 cases involved the right. The size of the lesions ranged from 6 to 80 mm (mean = 30).

RESULT

Mean surgery time was 90.8 minutes and mean hospital stay was 2.8 days. Two cases were converted to transperitoneal lateral approach due to the need to perform lymph node dissection. Two patients received blood transfusion.

CONCLUSION

Lateral approach retroperitoneoscopic adrenalectomy is safe and suitable for a wide range of adrenal pathologies and offers consistent clinical outcomes. When paracaval/paraaortic lymph node dissection is necessary, it can also be converted to a transperitoneal approach. Incidentalomas are common in the private sector due to ready access to axial imaging.

EP_A008

GERIATRIC ONSET OF PHEOCHROMOCYTOMA

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INTRODUCTION/BACKGROUND

Elderly pheochromocytoma (age ≥65 years) is a rare phenomenon, however, there have been increased detection rates because of advances in imaging and longer life expectancy. At Putrajaya Hospital, there was a total of 60 cases of pheochromocytoma between 2013 to 2023, with only 10% (6 cases) being in the elderly category.

CASE

We present two cases of pheochromocytoma in the elderly who presented as adrenal incidentalomas.

An 84-year-old Malay female with hypertension, type 2 diabetes mellitus(T2DM), and chronic kidney disease (CKD) stage 2 initially presented with palpitations, and was noted incidentally to have a right adrenal mass on abdominal ultrasound. Workup revealed elevated 24-hour urinary metanephrine 14.3 umol (0.33-1.53) and 24-hour urinary normetanephrine 4.6 umol (0.88-2.88). Adrenal CT revealed a well-defined right adrenal lesion measuring (3.5 x 3.2 x 3.4 cm) with plain HU +35. Patient underwent an uneventful open right adrenalectomy which revealed an adrenal tumour measuring 6.5 x 4 x 5 cm with capsular breach. She was discharged well after 6 days. Histopathology confirmed right adrenal pheochromocytoma with vascular and capsular invasion. Postoperatively, blood pressure was controlled on two antihypertensives with normal 24-hour urinary metanephrine and normetanephrine.

A 71-year-old Chinese female with T2DM, hypertension, and CKD stage 5 presented with incidental findings of a right adrenal mass on abdominal ultrasound during CKD workup. 24-hour urinary metanephrine revealed elevated urinary metanephrine 19.56 umol (0.33-1.53) and 24-hour urinary normetanephrine 55.66 umol (0.88-2.88).