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A CASE OF SYNCHRONOUS PANCREATIC NEUROENDOCRINE TUMOUR IN VON HIPPEL-LINDAU ASSOCIATED PHEOCHROMOCYTOMA

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Noor Ashikin Binti Ismail, Noor Lita Binti Adam, Nor Afidah Binti Karim
Hospital Tuanku Jaafar Seremban Negeri Sembilan, Malaysia

INTRODUCTION
Synchronous tumours in adrenal gland and pancreas are rare and considered as part of von Hippel-Lindau (VHL) syndrome. Because pancreatic neuroendocrine tumours (PNETs) in VHL have malignant potential, it is of clinical importance to diagnose these as early as possible. We report a case of synchronous PNETs in pheochromocytoma associated with VHL syndrome managed by surgical excision.

CASE
A 13-year-old male first presented to an ophthalmology clinic with bilateral eye floaters was found to have classic triad of catecholamines excess symptoms and severe hypertension. Family history was unremarkable except his maternal aunt had ovarian tumour. On examination, his blood pressure was 200/100 mmHg, regular pulse of 120 bpm, normal thyroid, abdominal, and neurological examinations. Fundus examination revealed grade IV hypertensive retinopathy. Electrocardiogram showed significantly increased left ventricular voltages. The diagnosis of clinically suspected pheochromocytoma was confirmed with 24-hour urine normetanephrine at 10 times upper limit normal. CECT adrenal showed lobulated lesion at right suprarenal measuring 5 x 3.6 x 5.4 cm with spontaneous density of 38 HU and absolute washout of 41%. Gallium-68 DOTATATE scan showed moderately avid right suprarenal mass suggestive of pheochromocytoma and intensely avid pancreatic lesion at head of pancreas (1.5 cm) suggestive of synchronous PNETs which was not detected on CT images. There was neither other localizations nor lymph nodes involvement.

He underwent right adrenalectomy and pancreatic nodulectomy with complete tumour excision. Genetic analysis revealed a missense mutation c.500G >A (p. Arg167Gln) in exon 3 of the VHL gene. On recent clinic review, he remained asymptomatic and normotensive without medication, and scheduled for follow-up urine metanephrines and MRI brain screening for hemangioblastomas.

CONCLUSION
Synchronous PNETs in VHL associated with pheochromocytoma are reportedly uncommon. Presence of the exon 3 mutation in VHL gene determines more aggressive course and metastases development of PNETs which requires early detection and intervention.

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TYPE 1 DIABETES AND LATENT AUTOIMMUNE DIABETES IN ADULTS: ARE THEY THE SAME?

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Low Yen Nee, Yoh Yee Yee, Goh Qing Ci, Tong Chin Voon
Department of Medicine, Hospital Melaka, Malaysia

INTRODUCTION
It is believed that LADA is the extreme end of T1DM as they shared similarities in terms of genetic susceptibility and presence of similar islet autoantibodies but presented in the older age.

METHODOLOGY
This is a cross-sectional study based in T1DM Clinic, Hospital Melaka where patients' information was collected via records and latest clinic visits. We aimed to compare T1DM and LADA patients in terms of phenotype, complications, co-morbidities and treatment.

RESULTS
We had 95 T1DM patients and 27 LADA patients in our clinic. LADA patients were older (54 ± 15 years old Vs 28 ± 17 years old, $P<0.001$) with later onset (43 ± 15 years old Vs 15 ± 10 years old, $P<0.001$) compared to T1DM patients.

Less LADA patients had diabetic ketoacidosis as the initial presentation but it is associated with higher rates of hypertension (44.4% vs 21.1%, $P=0.015$) and dyslipidemia (77.8% vs 36.8%, $P<0.001$). LADA patients had lower eGFR (90.4 ± 35 mls/min vs 111.0 ± 30.6 mls/min, $P=0.001$) and higher rate of ischemic heart disease (14.8% vs 3.2%, $P=0.042$).

Treatment wise, more LADA patients were on oral hypoglycemic agent (44.4% vs 20%, $P=0.01$), anti-hypertensives (59.3% vs 37.9%, $P=0.048$) and statins (81.5% vs 34.7%, $P<0.001$) but required less total daily dose of insulin per weight (0.60 ± 0.23 u/kg/day vs 0.75 ± 0.30 u/kg/day, $P=0.006$). There was no significant statistical difference in HbA1c between both groups.

CONCLUSION
Early recognition of LADA patients and intensive glycemic management in this group of patients may be crucial as they can present with more complications compared to T1DM patients.