PA-A-23

CHARACTERISTICS OF PATIENTS WITH TYPE 1 DM AND LADA IN A MALAYSIAN PUBLIC HOSPITAL: A CROSS- SECTIONAL STUDY

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INTRODUCTION
Type 1 diabetes mellitus (DM) occurs as the result of pancreatic beta cell destruction. Latent autoimmune diabetes in adults (LADA) is immunologically similar to T1DM but immune destruction progresses at a slower rate. The aim of the study is to identify the clinical characteristics of patients with T1DM and LADA in our clinic.

METHODOLOGY
This is a single centre cross-sectional study involving all 122 patients with T1DM and LADA. Information was obtained from patients' records and interviews during follow up.

RESULTS
There were 49 males (40.2%) and 73 females (59.8%) with a mean age of 35.3 (*SD* 14.9) years old. The mean duration of disease is 12.9 (*SD* 9.7) years. Ninety-five subjects (77.9%) have T1DM and 27 subjects (22.1%) have LADA. The most common complication was retinopathy (14.8%). Almost 2/3 of subjects (61.5%) reported having minor hypoglycemia and 15 (12.3%) had diabetic ketoacidosis in the past year. The most common co-morbid is dyslipidemia (45.9%). Eighteen percent of the subjects have other autoimmune diseases. Majority of the subjects were on at least one analogue insulin (93.4%) and on basal bolus regimen (89.3%). Only 6 subjects (4.9%) were on insulin pump. One hundred fourteen (93.4%) subjects performed self-monitoring of blood glucose (SMBG) and only 26 (21.3%) subjects have used continuous glucose monitoring systems (CGMS) at least once. The mean HbA1c is 8.91% (SD 2.2). The most frequent pancreatic autoantibodies detected were glutamic acid decarboxylase (GAD) (77.9%) and islet cell antibody (ICA) (77.7%).

CONCLUSION
Majority of our subjects with T1DM and LADA are on analogue insulin andon basal-bolus regimen with most of them performing SMBG. Despite this, the rate of hypoglycemia is high and control remains suboptimal. Increasing the use of technologies such as CGMS and insulin pumps which are not fully utilized at present, may improve outcomes.

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CASE REPORT: PRIMARY HYPERPARATHYROIDISM AND JAW TUMOUR SYNDROME WITH CDC73 GENE MUTATION IN A YOUNG PATIENT

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INTRODUCTION
Primary hyperparathyroidism (pHPT) occurs frequently in those over the age of 50 years. This condition is uncommon in young adults and are more likely to have an underlying germline mutation.

CASE
We present a case of a healthy 16-year-old male who was incidentally found to have an elevated calcium of 3.16 mmol/L. Family history revealed that his father, aunty and grandfather also had a history of hypercalcemia. No genetic study was done previously. The patient was diagnosed with iPTH-mediated hyperparathyroidism based on blood investigations. Localization scan revealed an overactive right parathyroid gland secreting excess iPTH. Subsequently, he was scheduled for right parathyroidectomy.

Histopathology report confirmed the diagnosis of a right superior parathyroid adenoma. His iPTH level declined from 968.5 pg/ml to 7.9 pg/ml after the surgery while calcium and ALP levels also normalized. He subsequently required calcium and activated Vitamin D supplementation.

The patient and his family were referred for further genetic assessment, revealing CDC73-related disorders, with a pathogenic mutation on CDC73 gene. The patient's father was found to develop a jaw tumour with histologic confirmation of invasive ossifying fibroma. Hence, tumour debulking was planned.

CDC73-related disorder is an autosomal dominant disorder resulting from the inactivation of the CDC73 tumor suppressor gene. The spectrum includes: Hyperparathyroidism jaw tumor (HPT-JT) syndrome, parathyroid carcinoma and familial isolated hyperparathyroidism (FIHP). Penetration of pHPT is as high as 80% to 95%, while parathyroid carcinoma may be found in more than 20% of patients. Lifelong surveillance is indicated for positive gene carriers to look for recurrent hyperparathyroidism, parathyroid carcinoma, renal and uterine tumour in females.

CONCLUSION

This case report demonstrated the importance of early detection of hypercalcaemia in a younger population, the need to obtain a comprehensive family history and the significance of proceeding with early germline CDC73 mutation detection for optimal clinical management of pHPT. Genetic counselling and surveillance of family members who are at risk of developing CDC73-related disorders must also be emphasized.

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MICROANGIOPATHIC HEMOLYTIC ANEMIA IN A CASE OF PHEOCHROMOCYTOMA CRISIS

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INTRODUCTION

Pheochromocytomas are rare neuroendocrine tumours of the adrenal medulla. Pheochromocytoma crisis, a potentially fatal presentation, is characterized by severe hypertension or hypotension with end organ damage occurring spontaneously or precipitated by a stressor such as surgery, stress or medications such as anaesthetic agents, corticosteroids, and metoclopramide. A cascade of life threatening events may occur including myocardial infarction, acute pulmonary oedema, stroke, paralytic ileus, thrombosis, renal failure, diabetic ketoacidosis, and lactic acidosis. Microangiopathic hemolytic anemia (MAHA), an uncommon sequelae of pheochromocytoma crisis or malignant hypertension can occur.

CASE

We report a case of pheochromocytoma crisis presenting with microangiopathic hemolytic anemia. A 58-year-old female with diabetes mellitus, hypertension, and dyslipidemia presented with recurrent vomiting and epigastric pain for 9 days. On arrival, her blood pressure was 177/133 mmHg with a pulse rate of 146 beats/minute. It worsened to 214/146 mmHg following the administration of intravenous metoclopramide. She developed acute kidney injury (AKI) (urea 14.5 mmol/L, creatinine 321.8 umol/L) and MAHA with Coomb's test negative anemia. Her hemoglobin decreased from 10.6 g/dl to 8.1 g/dl in 2 days, with increased schistocytes from 2.7% to 3.0%, accompanied by reticulocytosis of 4.8%, thrombocytopenia with platelet of 141 k, and raised lactate dehydrogenase of 482 U/L. She received 1-unit packed cell transfusion. Her MAHA and her AKI resolved subsequently following adequate blood pressure control with alpha blockade. Twenty four hour urine metanephrine was elevated with abdominal imaging demonstrated a right adrenal mass (4.0x 4.5x 4.0 cm) supporting the diagnosis of a right adrenal pheochromocytoma. She underwent a right open adrenalectomy with symptoms resolution and normotension after surgery.

CONCLUSION

This case highlights the awareness of detecting a pheochromocytoma crisis and avoiding medication that may induce a pheochromocytoma crisis. High index of suspicion, prompt diagnosis and early initiation of treatment of pheochromocytoma crisis is essential to minimise complications and improve clinical outcomes.