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A CASE OF CHILDHOOD ADRENOCORTICAL TUMOR: A TICKING TIME BOMB

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INTRODUCTION/BACKGROUND

Childhood adrenocortical tumours (ACTs) are rare. Majority of children have clinical evidence of hormone hypersecretion at diagnosis which varies from virilization to Cushing's syndrome. Markers for risk stratification in ACTs are not well-defined and differentiating adenomas from carcinomas remain challenging.

CASE

We describe a child with adrenocortical tumour presenting with virilization and hypertensive crisis.

A 6-year-old male was initially admitted for status epilepticus attributed to rotavirus acute gastroenteritis. He had hirsutism, pubic hair, penile enlargement, and gynecomastia, which started 2 years ago. His weight and height were at the 50th centile. He developed hypertensive crisis requiring five anti-hypertensive agents including parenteral infusion. Blood tests showed high testosterone, renin and aldosterone. 24-hour urinary sample demonstrated hypercortisolism. Magnetic resonance imaging of the upper abdomen showed a left adrenal mass. He underwent complete resection of the solid mass, which measured 83 x 82 x 59 mm and showed no signs of infiltration. The tumour scored 2/9 using the Wieneke algorithm, features compatible with adrenocortical adenoma. Post-resection, his blood pressure normalized with a single oral anti-hypertensive. He was counselled for adjuvant chemotherapy, but the family opted for watchful management.

CONCLUSION

The paucity of data on prognostic factors, disease staging, and treatment hinders the clinician's ability in identifying patients at high risk of relapse even after complete tumour resection. More targeted predictive models for recurrence or malignant risk will be helpful in guiding treatment strategies.

EP_P013

CONGENITAL HYPOTHYROIDISM IN A TWIN MISSED BY NEWBORN SCREENING: A CASE REPORT

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INTRODUCTION/BACKGROUND

Thyroid hormone plays a significant role in the development of multiple organ systems especially the brain. Newborn screening program enables early detection and treatment of congenital hypothyroidism (CH) to prevent long-term cognitive and physical impairment. We report a case of a twin with CH missed by newborn screening using umbilical cord blood thyroid-stimulating hormone (TSH) in Malaysia.

CASE

A 13-year-old Malay female was born at 34 weeks of gestation with a birth weight of 1600 g. Her cord TSH screening was normal at birth (6.54 mIU/L). She is the first twin of monozygotic monoamniotic twins. Her larger twin sister weighed 2470 g at birth and also had normal cord TSH (3.12 mIU/L). The patient was admitted at birth for feeding establishment, nosocomial infection and neonatal jaundice. She presented later at 2 months of age with abdominal distension, constipation and lethargy, and was treated for sepsis. At 5 months of age, she was noted to have faltering growth, developmental delay, and constipation. Her growth parameters were way below the 3rd percentile (weight 3400 g, length 53 cm and head circumference of 34 cm) and she also had macroglossia and hypotonia. Thyroid function test (TFT) showed severe primary hypothyroidism (TSH 194 uIU/mL, free T4 <5.15 pmol/L). She was initiated on oral thyroxine 15 mcg/kg/day at 5 months of age and her TFTs normalized within a month of treatment. Ultrasound of the thyroid gland at 3 years of age revealed thyroid gland hypoplasia. Her twin had normal growth and development.

CONCLUSION

This case illustrates the need to retest the thyroid function in same-sex twins even when the cord TSH screening is normal. There should also be a high index of suspicion on the diagnosis of congenital hypothyroidism in infants with faltering growth, constipation, or developmental delay.