HPA axis secondary to exogenous steroids. Renin (84.80 mU/L), aldosterone (280.40 pmol/L), testosterone (0.31 nmol/L), estradiol (85.9 pmol/L) and thyroid function were normal. Physiological oral hydrocortisone replacement was promptly commenced, with education on adrenal crisis and sick days while awaiting recovery of the HPA axis.

CONCLUSION

This case illustrates the serious complication of Cushing syndrome with secondary HPA axis suppression due to repeated intralesional TAC therapy for paediatric keloid scars. Factors such as appropriate paediatric doses, size of the keloid, site of injection (dermal, not subcutaneous), and frequency of injections should all be given due consideration before using TAC. Clinicians prescribing steroids to children should be aware that suppression of the HPA axis is a very real and life-threatening complication and that steroids should be prescribed judiciously.

EP_P021

GROWTH HORMONE THERAPY IN PAEDIATRIC ONCOLOGY SURVIVORS: CASE SERIES FROM A MALAYSIAN TERTIARY CENTRE

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INTRODUCTION

Growth hormone deficiency (GHD) is a recognized endocrine complication among childhood cancer survivors resulting from disruption of hypothalamic–pituitary axis due to tumors, neurosurgery, or cranial irradiation. While recombinant human growth hormone (rhGH) therapy improves growth and metabolic outcome, concerns remain regarding its long-term safety with risk of tumor recurrence and secondary neoplasm. We present a case series of five paediatric oncology survivors with confirmed GHD receiving rhGH in Hospital Tunku Azizah, highlighting our clinical experience in comparison with international practice.

CASE

Five paediatric oncology survivors (acute lymphoblastic leukemia, craniopharyngioma, medulloblastoma, and supratentorial PNET) with GHD were commenced on rhGH therapy 2.5–5.5 years after completion of cancer treatment. All patients had significant short stature with a mean height SDS of −3.22 prior to the initiation of therapy. GHD was confirmed biochemically with low IGF-1 level and dynamic testing (peak GH 0.35–4.61 ng/mL). Among the four patients with brain tumors, two had stable residual disease while the remaining were tumor-free. rhGH therapy was temporarily ceased in two patients due to minor increment in tumor size but was successfully resumed following stabilization without further complications to date. Two patients are concurrently on pubertal induction. Most patients demonstrated catch-up growth with increased height velocity, supporting the efficacy of rhGH in this population. No secondary malignancies or significant adverse events were observed during follow-up.

CONCLUSION

Paediatric oncology survivors with GHD in this series demonstrated a favorable response to rhGH therapy, evidenced by improvements in height SDS and growth velocity, while maintaining oncological stability in the majority of cases. Our findings are consistent with international data, supporting cautious but appropriate use of rhGH in this population. Careful patient selection and close multidisciplinary monitoring remain essential.

EP_P022

WHEN PUBERTY COMES TOO SOON: CENTRAL PRECOCIOUS PUBERTY WITH PITUITARY MICROADENOMA AND PINEAL CYST

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INTRODUCTION

Central precocious puberty (CPP) is defined as premature activation of the hypothalamic-pituitary-gonadal axis, resulting in the development of secondary sexual characteristics before 8 years of age in girls. While most cases are idiopathic, intracranial pathologies such as pituitary lesions may cause CPP. Early identification is essential to optimize outcomes and minimize psychosocial impact. We report a case of CPP associated with pituitary microadenoma and pineal cyst from the Paediatric Department, Hospital Sultanah Nur Zahirah.

CASE

A 4-year-old female presented with bilateral breast enlargement, with ultrasonography showing normal fibroglandular tissue. Breast development was noted since 3 months of age, with whitish vaginal discharge from 2 years. She demonstrated accelerated growth, with height and weight above the 95th centile. There were no features suggestive of peripheral causes or raised intracranial pressure. Examination revealed Tanner stage A1B3P2. Bone age was markedly advanced (11 years vs. chronological age 4 years).

Hormonal evaluation showed elevated LH (3.28 IU/L) and follicle-stimulating hormone (4.79 IU/L), consistent with CPP. MRI demonstrated focal delayed enhancement in the posterolateral pituitary suggestive of microadenoma, and an incidental pineal cyst without complex features. Ophthalmological assessment was normal. Neurosurgical evaluation advised conservative management with annual imaging surveillance. Intramuscular Triptorelin was initiated at 4 years 1 month. Follow-up showed regression of pubertal progression, resolution of vaginal discharge, and reduced growth velocity to 6 cm/year.

CONCLUSION

This case highlights the importance of thorough evaluation in CPP to identify underlying intracranial pathology. Early initiation of gonadotropin-releasing hormone analog therapy effectively halts pubertal progression and preserves growth potential.

EP_P023

BECKWITH-WIEDEMANN SYNDROME PRESENTING AS PERSISTENT NON-KETOTIC HYPOGLYCEMIA IN A PRETERM INFANT

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INTRODUCTION

Beckwith–Wiedemann syndrome (BWS) is a congenital overgrowth disorder associated with dysregulation of genes on chromosome 11p15.5. Infants with BWS can present with hyperinsulinemic hypoglycemia. There are also distinctive clinical features including macrosomia, macroglossia and visceromegaly that may raise suspicion of this diagnosis.

CASE

A preterm female infant of 30 weeks gestation with a birth weight of 1.87 kg (97th centile) had recurrent hypoglycemia in the neonatal period, requiring escalation to a maximum glucose infusion rate of 17.6 mg/kg/min. Hypoglycemia only resolved after starting intravenous glucagon infusion. Critical sampling during hypoglycemia revealed serum insulin 3.9 µU/mL (3–25 µU/mL), serum ketone 0.2 mmol/L, cortisol 3,053 nmol/L and growth hormone 16.2 ng/mL. These findings supported the diagnosis of non-ketotic hyperinsulinemic hypoglycemia. She was commenced on oral diazoxide with resolution of hypoglycemia and discontinuation of glucagon.

Examination at birth revealed macroglossia, hepatomegaly and ballotable kidneys. An initial US abdomen at birth revealed bilateral enlarged kidneys but normal liver. An initial chromosomal study revealed karyotype 46, XX. Over time, additional clinical features became evident fulfilling the diagnostic criteria for BWS, that is, polyhydramnios, large for gestational age, transient hypoglycemia, hyperinsulinism, macroglossia, hemihypertrophy, facial nevi, bilateral ear creases, hepatomegaly and ballotable kidney.

A repeat US abdomen surveillance at 4 months old revealed a heterogeneous liver mass with marked vascularity, prompting a diagnosis of hepatic hemangioma. She was commenced on oral propranolol, with reduction in the size and vascularity of the liver hemangioma and decline in alpha-fetoprotein levels.

CONCLUSION

The clinical features of BWS may not be recognizable in a preterm baby in early neonatal period. Careful clinical examination should be done in a baby with persistent hyperinsulinemic hypoglycemia for underlying syndromal causes. US surveillance should also be carried out due to increased risk of hepatoblastoma or liver hemangioma in BWS, as was seen in this case.

EP_P024

BEYOND OBESITY: DIAGNOSTIC CHALLENGES OF BARDET-BIEDL SYNDROME

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INTRODUCTION

Bardet-Biedl Syndrome is a rare autosomal recessive disorder characterized by multisystem manifestations, such as early-onset obesity, hypogonadotropic hypogonadism, retinal dystrophy, polydactyly, renal anomalies and intellectual disability. Phenotypic variability makes diagnosis challenging. Central obesity and hypogonadotropic hypogonadism are frequently observed, but often overlooked.

CASE

We report a 12-year-old male who was referred at the age of 10 years for evaluation of central obesity and buried penis. He was born at term with birth weight of 3,200 g. Both parents were non-consanguineous, and antenatal history was unremarkable. Early-onset excessive weight gain was noted from infancy, with hyperphagia during early childhood.