Paediatrics E-Poster

EP_P001

A CHILD WITH AN AGGRESSIVE FUNCTIONAL ADRENOCORTICAL CARCINOMA

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INTRODUCTION
Adrenocortical carcinomas, among the rarest and most aggressive paediatric endocrine tumours, manifest with diverse symptoms like virilization, Cushing's syndrome, or both.

CASE
We present a case of functional adrenocortical carcinoma in a female aged 6 years and 7 months, who initially presented with hypertensive encephalopathy and hypokalaemic hypochloremic metabolic alkalosis, which resolved with symptomatic treatments. Ten months later, she presented with frank Cushing's syndrome, refractory hypertension, generalised virilization, extensive skin fungal infection and severe backache. Breast cancer was diagnosed in her maternal aunt. Hormonal tests showed non-ACTH dependent hypercortisolism and marked elevation of androgens. Computed tomography revealed a large left suprarenal mass, with multi-focal liver lesions and lung nodules suggestive of distant metastasis, left renal vein thrombosis and multiple osteoporotic vertebral fractures.
A clinical diagnosis of stage 4 functional adrenocortical carcinoma was made. While complete surgical removal of the tumour is the gold standard, it was not feasible immediately due to the substantial size of the tumour and presence of distant metastases. Neo-adjuvant chemotherapy was started. Mitotane and ketoconazole were introduced concomitantly to control hypercortisolism, with initial success. Hydrocortisone replacement was needed for a short period when there was a rapid decline in cortisol levels following chemotherapy. Unfortunately, with poor commitment from the family, the disease advanced rapidly with worsening lung and liver metastases. Following a family conference, the parents opted for palliative treatment with mitotane monotherapy, and the child was transferred to the district hospital for comfort care.

CONCLUSION
Medical treatment is useful in controlling the symptoms of severe hypercortisolism. Steroid replacement may be needed with the use of adrenolytic agent. Adrenocortical carcinoma is aggressive and a high index of suspicion is needed for early diagnosis.

EP_P002

CONGENITAL HYPERINSULINISM SECONDARY TO ABCC8 MUTATION: A CASE STUDY

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INTRODUCTION
Congenital hyperinsulinism (CHI) results in persistent hypoglycaemia beyond infancy. Mutations in the ABCC8 and KCNJ11 genes are the most common aetiologies of congenital hyperinsulinism that leads to inappropriate insulin secretion irrespective of hypoglycaemia.

CASE
A 3-month-old male was referred to the clinic for persistent hypoglycaemia. He was born with a birth weight of 4480 g by elective caesarean section for macrosomia. His mother had an uneventful antenatal period and had a normal OGTT during the pregnancy.

Post-delivery, he was initially well until when he developed hypoglycaemia at 28 hours of life. He was transferred to the NICU from the postnatal ward. Hypoglycaemia was persistent requiring high glucose delivery rate up to 10 mg/kg/min. Glucagon infusion was started and was difficult to wean. Hypoglycaemic hyperinsulinaemia was confirmed at day 21 of life, with concomitant results of insulin 3.4 m IU/L and random blood glucose 1.8 mmol/L. Diazoxide was started. Glucagon infusion was stopped at day 26 of life, and the baby was discharged.