PE-23

VIRILISATION VS NON VIRILISATION: MULTIFACES OF CHILDHOOD ADRENOCORTICAL CARCINOMA (ACC)

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INTRODUCTION

Adrenocortical carcinoma (ACC) is a rare tumour in children. Unlike adults where ACC are non-functional (40%), the majority of tumours in children (94%) are functional. Their presentation varies from virilisation to Cushing syndrome. Early recognition is imperative in their management.

We describe 2 children with ACC with different clinical presentations.

Case 1: A 2-year-old girl presented with signs of progressive virilisation and abdominal distension for 5 months. Serial growth parameters showed rapid weight gain (crossed from the 3rd to 25th centile) although her height remained at 3rd percentile. She was normotensive. There were deepening of voice, hirsutism, clitoromegaly and pubic hair. There was an irregular hard mass at the left hypochondrium. Blood test showed hyperandrogenism and hypercortisolism. CT scan showed a left adrenal mass with bilateral lung metastases. She underwent resection of the adrenal mass followed by bilateral lung nodulectomies. Histopathological examination confirmed ACC with lung metastases. She had completed concurrent mitotane and chemotherapy.

Case 2: A 6-year-old boy presented with hypertensive encephalopathy preceded by rapid weight gain. There were hirsutism and acanthosis nigricans. Ultrasound imaging showed a left adrenal mass, confirmed by CT scan. There was no metastatic disease. Blood test showed hypercortisolism. Complete tumour resection was achieved. Histopathological examination confirmed ACC.

CONCLUSION

ACC is potentially fatal. Since ACC in children is functional, it is hoped that increased familiarity with its presentation will result in earlier diagnosis, intervention and improvement in their overall survival.

PE-24

HYPERINSULINAEMIC HYPOGLYCAEMIA (HH) IN A MOSAIC TURNER SYNDROME TODDLER

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INTRODUCTION

Hyperinsulinaemic hypoglycaemia(HH) is a rare but important cause of hypoglycaemia, especially in the newborn. Early identification and diligent management of these patients is vital to prevent neurological insult. We report an interesting case of a toddler with mosaic Turner Syndrome(TS) with HH, responsive to diazoxide treatment.

RESULTS

A 25-month-old girl was born term via Emergency Lower Segment Caesarean Section(EMLSCS) for fetal distress with a birth weight 2.75 kg. No significant antenatal issue noted. Her Apgar score were 9 and 10 at 1 and 5 minutes of life. However, she was admitted to Neonatal Intensive Care Unit(NICU) at 7 hours of life, following symptomatic hypoglycaemia, whereby her capillary blood glucose recorded was 0.9 mmol/L. She required initial D10% bolus 3 mls/kg and subsequently required maintenance dextrose intravenous infusion with highest Glucose Delivery Rate of 14.4mg/kg/hour to maintain normoglycaemia. Despite that, there were recurrent episodes of hypoglycaemia. She was started on IVI Glucagon on day 9 of life, with highest concentration of 20 mcg/kg/hour. Oral Diazoxide was started at day 17 of life following measurable insulin level during critical sampling when the child developed significant hypoglycaemia. Currently, she is still on oral Diazoxide 2.6 mg/kg/day with no hypoglycaemic episodes. Karyotype was sent in view of subtle features of TS which includes high arch palate and hypertelorism and revealed Mosaic TS with 46X, +mar (18)/45,X(12).

CONCLUSION

In summary, we report an interesting association of mosaic Turner Syndrome with Hyperinsulinaemic Hypoglycaemia. The recognition of hypoglycaemia in this group of patients is vital, as untreated hypoglycaemia may lead to irreversible brain damage. The mechanism leading to hyperinsulinism in this condition is not well established to the best of our knowledge and warrants further research.