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ABSTRACTS
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Ectopic ACTH-secreting Primitive Neuroectodermal Tumour (PNET) of the sympathetic chain in a boy with Cushing's syndrome

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Endogenous Cushing's syndrome (CS) in children is rare. Most of these patients had Cushing's syndrome due to ACTH-independent adrenal tumours, rarely from ACTH-producing pituitary tumour, even rarely from an ectopic ACTH-producing tumour. We report a rare ACTH-secreting primitive neuroectodermal tumour (PNET) of the sympathetic chain in a boy with Cushing's syndrome. An 11-year-old boy presented with excessive weight gain and masculinisation for 3 months. There were also intermittent blurring of vision and fatigue. Physical examination revealed a pigmented, masculinised obese boy (BMI= 28.5kg/m2) with acne, hypertension (BP 150/100 mmHg) and glaucoma. Serum assays confirmed hypercortisolism with loss of diurnal variation (8AM serum cortisol was 996nmol/L, midnight was 1260nmol/L (N= 85-460nmol/L). His 24-hour urinary free cortisol was 13,299nmol/24hr (N= 8-590nmol/L) and high serum ACTH, 264pg/ml (N= 0-46pg/ml). Testosterone was 4.1nmol/L (N=8.4-28.7nmol/L), while potassium was 2.5mmol/L. Overnight high-dose dexamethasone suppression test failed to suppress serum cortisol. Pituitary magnetic resonance imaging (MRI) was inconclusive. Whole body computerized tomography showed a large intra-abdominal mass likely arising from the left adrenal gland. His condition was complicated with hypertensive encephalopathy, cardiac failure and ocular hypertension, all were treated symptomatically. Intraoperatively a tumour from sympathetic chain was removed. Histopathological examination, immunophenotyping and immunohistochemistry confirmed PNET. Postoperatively serum cortisol and intra-ocular pressure were normal but hypertension remained. Ectopic ACTH-producing tumour should be suspected in patients with Cushing's syndrome who are pigmented, with severe complications of hypercortisolism which fails to be suppressed by high dose dexamethasone test. Symptomatic treatment of complications should be aggressive and prompt surgical removal of tumour done after its localization.