Case reports

Cushing syndrome due to an adrenal phaeochromocytoma

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Introduction

Adrenal phaeochromocytoma is a rare cause of Cushing syndrome [1–3]. We describe a 16-year old girl with Cushing syndrome with a right suprarenal mass on CT scanning which was histologically confirmed.

Case report

A 16-year old schoolgirl with rapid onset of weight gain had central obesity, purplish streaky lesions in the upper thighs, abdomen and buttocks, hair loss and acne. She had amenorrhoea throughout the period and also noticed progressive difficulty in rising from the squatting position. On examination her BMI was 22.8. She was plethoric, had a moon face, a buffalo hump, frontal hair loss (Figure 1) and central obesity with purple striae over the abdomen, thighs and calves. She had a fine tremor, some hyperpigmented patches in both hands and grade 4 proximal muscle weakness without wasting, with normal tendon reflexes. Her blood pressure was 160/110 mmHg without a postural drop. There was no hirsutism or signs of virilisation.

The low dose dexamethasone suppression test was positive [basal cortisol level was over 2000 nmol/L and unchanged after 48 hours of dexamethasone]. After high dose dexamethasone the serum cortisol level was still over 2000 nmol/L. Ultrasound scan of the abdomen showed a round solid mass with mixed echogenicity (8 cm × 8.9 cm) in the right suprarenal area, and CT scan confirmed a mixed density right suprarenal mass (Figure 2). Both scans suggested a phaeochromocytoma, and not a cortical tumour. Urinary VMA, metanephrines and plasma ACTH were not performed. She underwent an uneventful right adrenalectomy and the histology of the tumour (Figure 3) showed a phaeochromocytoma

Figure 1. Facial features of Cushing syndrome in the patient.

Figure 2. CT scan of the abdomen showing the adrenal medullary tumor.

Figure 3. A histological section of the adrenal medullary tumour showing poor prognostic features.
with poor prognostic features with pleomorphism, tumour necrosis high mitosis. Post-operatively she recovered well and was discharged on replacement steroids, subsequently tailed off.

Comment

Adrenal medullary phaeochromocytomas could rarely cause Cushing syndrome by causing ectopic ACTH hormone secretion [4,5]. Ignoring this possibility could lead to severe peri-operative complications when resection of the tumor is envisaged.

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References


Herniation of a hamartomatous growth of the liver in an infant masquerading as a cardiac tumour

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Introduction

Congenital diaphragmatic hernia is a common birth defect. The incidence varies from 1 in 5000 live births to 1 in 2000, if stillbirths are included. Herniation through the diaphragm could occur through the oesophageal hiatus, the left or right foramen of Bochdalek, the foramen of Morgagni, and communications caused by congenital absence of the left diaphragm or the central tendon [2]. They commonly occur on the left side (75–80%) and occasionally bilaterally [1]. We report an unusual right-sided diaphragmatic hernia, occurring through the central tendon which was initially misdiagnosed as a cardiac tumour.

Case history

A baby girl was born vaginally to non-consanguineous parents as 38 weeks of gestation. Birthweight was 2.3 kg. There was no perinatal complication but she had dysmorphic features comprising microcephaly (OFC 31cm; <3rd centile; NCHS 2000), polydactyly, cutis aplasia and low set abnormal ears. The cardiovascular system and abdomen were clinically normal. She developed jaundice and shortness of breath on the third day and was admitted to hospital. Serum bilirubin was below phototherapy level and other investigations excluded sepsis. 2D echocardiography showed hypertrophy of both ventricles with mild systolic dysfunction. Based on echocardiography a diagnosis of mild infiltrative cardiomyopathy was made. She was discharged at the age of 7 days, awaiting the report on karyotyping (karyotyping was normal).

At 10 weeks of age, the child was readmitted with the complaints of peripheral cyanosis and dyspnoea. She was restless. The weight was 3.5 kg; OFC was 34 cm (<3rd centile; NCHS 2000). Early clubbing was present. A hypopigmented skin lesion was noted on the chest. Cardiovascular system was clinically normal. Repeat echocardiography revealed a small ostium secundum ASD. Biventricular hypertrophy and a moderate sized pericardial effusion was seen. A large tumour like mass was seen on the right side of the pericardial cavity, measuring about 3.5 cm × 2.5 cm. CT scan of the thorax was done and a mass lesion was confirmed. While in the ward the child developed generalised tonic seizures. A clinical diagnosis of tuberous sclerosis (TS) was made on the basis of seizures and the hypopigmented skin lesions. The cardiac tumour was presumed to be a

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