PHOTO ASSAY

Organic Hypercortisolism of Childhood: Adrenal Cushing's Syndrome

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Abstract

Adrenocortical tumors (ACTs) only account for 0.3–0.4% of all neoplasms in childhood. In this article, we have discussed the clinical features, evaluation, and treatment of adrenal Cushing's syndrome (CS) and the features which distinguish it from adult CS.

Keywords: Adrenal adenoma, Hypercortisolism of childhood, Pediatric Cushing's syndrome.

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Case Description

The picture is of a 12-month-old child who was brought to the hospital by his parents with complaints of excessive weight gain over the last 4 months associated with poor appetite, sleeplessness, and irritability (Fig. 1).

He was diagnosed to have CS secondary to a mixed-secreting left adrenal tumor which was reported as Adrenocortical carcinoma on histopathological examination, Wieneke score – 3 (borderline) (Fig. 2).

We present this child with the classical features of CS due to a mixed tumor (cortisol and testosterone secreting adenoma) normalizing within 5 days of laparoscopic adrenalectomy and also the feasibility of laparoscopic adrenalectomy in a child under the age of 1 year (Figs 3 and 4).

Discussion

Cushing's syndrome is a rare entity in children with the incidence being 2–5/10,00,000 per year. The most common cause of CS in children is iatrogenic (exogenous administration).[1]

Adrenocortical tumors are rare in childhood accounting for only 0.3–0.4% of all neoplasms in this age group. Yet, in children less than 7 years of age, adrenal pathologies [adenoma, carcinoma, adrenocorticotropic hormone (ACTH) independent adrenal macronodular hyperplasia, and primary pigmented nodular hyperplasia] are the most common causes of CS whereas in those more than 7 years old, Cushing's disease constitutes the maximum chunk of cases.[2]

Fig. 1: Clinical features of CS in a child

The most common presenting feature of CS in children that distinguishes them from adults is the ***"linear height deceleration with concomitant weight gain."*** Height and body mass index (BMI) standard deviation scores (SDS) help differentiate children with CS from those with simple obesity.[1]

Other signs noted in pediatric Cushing's are facial plethora, lanugo-like hair, hirsutism, moon facies, supratemporal and supraclavicular fat pads, central obesity, violaceous striae over trunk, sleeplessness, irritability, fatigue, and hypertension.[2]

As in adults, to establish the diagnosis of Cushing's, hypercortisolism should be documented by screening first, either by 24-hour urinary free cortisol or late-night salivary cortisol followed by confirmation by a low-dose dexamethasone-suppression test.

Once the diagnosis of Cushing syndrome is confirmed, investigations to distinguish ACTH-dependent disease from the

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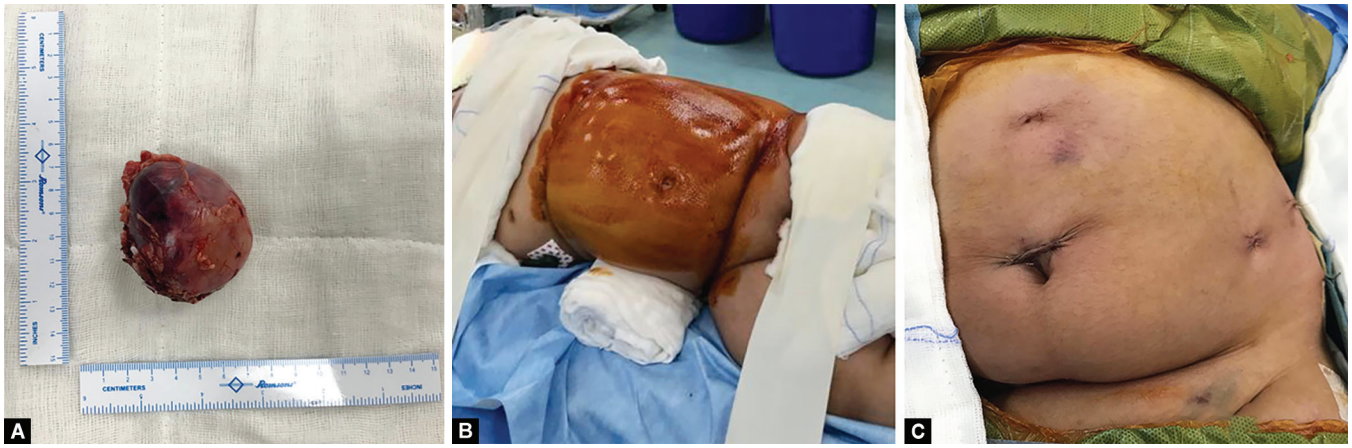

Figs 2A to C: (A) Specimen of mixed hormone secreting left adrenocortical carcinoma; (B) Patient positioning for laparoscopic left adrenalectomy; (C) After closure of port site scars

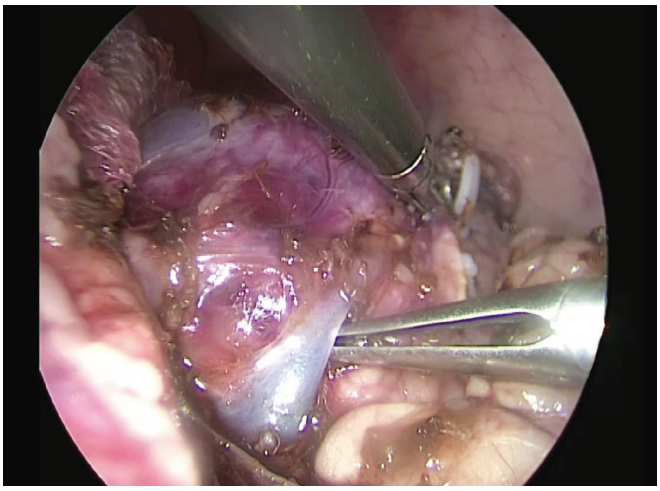

Fig. 3: Left adrenal vein identification and ligation during laparoscopic adrenalectomy

ACTH-independent disease should be done. Surgical intervention is the first line of treatment for such adrenal lesions.[1]

Children with adrenocortical carcinomas usually have germline p53 mutations. The prognosis of adrenocortical carcinoma in children is very variable and difficult to predict in clinical practice, with the 5-year survival rate being 30–70% and below 20% in metastatic disease.[3]

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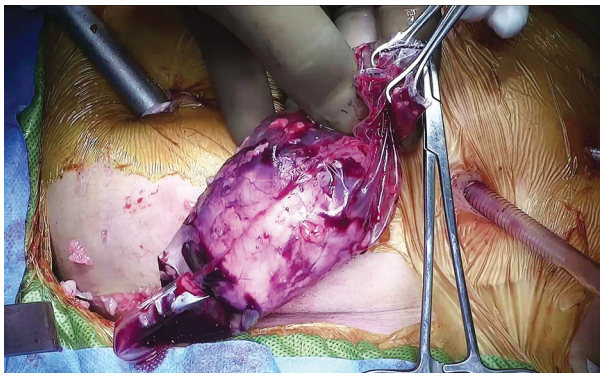

Fig. 4: Specimen retrieval in an EndoBag *via* Pfannenstiel incision

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