Case Report

Adrenocorticotropic Hormone Independent Adrenal Adenoma in Children a Rarity: Report of Two Cases and Review of Literature

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Abstract

Adrenocortical adenoma is a rare tumour in children and even rarer in infancy. Most of these tumours in the paediatric age group are hormonally active and predominantly present with virilisation. Cortisol hypersecretion, presenting as Cushing syndrome, is extremely rare and seen in older age groups. We are reporting two cases of adrenal adenoma with different clinical presentations. The first case was of a 9-month-old female who presented with isolated features of Cushing's syndrome (CS). The second case was of a 3-year-old male who presented with features of CS, virilisation and dyselectrolytemia. The clinical, biochemical and histological features are discussed. These cases are being reported because of their rarity. Early diagnosis, proper peri-operative management, complete excision of the tumour and close follow-up are crucial to improve survival and quality of life.

Keywords: Adenoma, adrenocortical tumour, Cushing's syndrome, dyselectrolytemia, infant, paediatric, virilisation

Introduction

Adrenocortical tumour (ACT) is a very rare malignancy in children. It comprises 0.2% of the paediatric cancers.[1] In the United States, <25 new cases of ACT are diagnosed annually with an estimated annual incidence of 0.2–0.3 cases per 1 million individuals.[2] The typical age of presentation is <5 years with a median age of 3–4 years.[3] Most common presentation (80%–90%) is secondary to hormonal excess with isolated virilisation or a combination of hypercortisolism and virilisation. The largest reported series of 254 children with ACTs suggested that 55% of patients presented with virilisation alone, and isolated Cushing's syndrome (CS) was found in only 5.5% of patients.[4] Complete surgical excision is the mainstay of treatment. Even after complete surgical excision, the recurrence rate was 23% to 85% of patients.[5] We wish to present two cases of adrenal tumour with different clinical presentations.

Case Reports

Case 1

A 9-month-old female infant presented to our outpatient department with a history of progressive weight gain and facial swelling for 6–7 months. There was no history of fever, vomiting or rashes over the body. Bladder and bowel habits were normal. She was delivered by normal vaginal route at the 38th week of gestation. Her birth weight was 3 kg and immunised as per the national immunisation schedule.

At initial presentation weight of the child was 11 kg. On examination, vital signs including heart rate (HR), blood pressure (BP) and respiratory rate (RR) were within normal limits. On physical examination, she had Cushingoid facies, buffalo hump and generalised excessive subcutaneous fat. There were no signs of facial acne, virilisation or striae over the body [Figure 1a].

Laboratory evaluation revealed that serum cortisol was 25.5 µg/dl (normal value 4.5–22.7 µg/dl), dehydroepiandrosterone sulphate 119 µg/dl (normal value 16–96 µg/dl) and serum adrenocorticotropic hormone (ACTH) was <5 pg/ml (normal value 10–60 pg/ml). Other biochemical investigations were within normal limits (Na–135 mEq/l,

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K–3.8 mEq/l and Cl–104 mEq/l). Abdominal ultrasound showed a well-defined solid hypoechoic mass measuring 2.8 cm × 2.1 cm × 2.5 cm in size in the region of the left adrenal gland. Abdominal contrast-enhanced computed tomography (CECT) showed that a well-defined soft tissue density mass measuring 2.2 cm × 2.4 cm × 2.1 cm at the left suprarenal region likely arising from the left adrenal gland [Figure 2a]. Mild heterogeneous enhancement was seen in post-contrast with an absolute percentage washout is 41.9%. The mass was abutting the spleen superiorly and the tail of the pancreas anterosuperiorly with preserved intervening fat plane. We planned for an exploratory laparotomy. On exploration with left subcostal incision, left adrenalectomy was done. Intraoperative findings were a 3.0 cm × 3.5 cm well-encapsulated mass arising from the left adrenal [Figure 2b]. Liver, spleen, pancreas and left kidney appeared grossly normal. The tumour was resected completely without breach of the capsule. Gross pathological examination revealed a well-encapsulated pale brown globular mass of size 3 cm × 3 cm × 2.5 cm and weight 9 g. On the cut section, a solid yellow area and a single whitish nodular area of size 1.5 cm were identified. Microscopic examination revealed that the well-circumscribed mass was arranged in small nests, cords and diffuse sheets. The tumour consisted of abundant eosinophilic cytoplasm with round nuclei with vesicular to irregular clumped chromatin. There was no atypical mitosis, necrosis or vascular invasion. Histopathological features were suggestive of an adrenocortical adenoma.

After surgery, the baby was treated with stress dosing of hydrocortisone. The post-operative period was uneventful, and discharged on maintenance doses of hydrocortisone which were subsequently weaned off. No adjuvant treatment was given post-operatively. During the 2-year follow-up period, her linear growth velocity improved and her weight normalised [Figure 1b].

Case 2

A 3-year-old male child was admitted under the paediatric department with a history of progressive weight gain and facial swelling for 2 years. He had a history of generalised tonic–clonic seizures for 6 6-month duration and was on anticonvulsant medication. Bladder and bowel habits were normal. He was born to non-consanguineous parents at the 38th weeks of gestation by normal vaginal route. His birth weight was 2.5 kg and immunised as per age.

At the time of admission, his weight was 20 kg (>97th percentile), length 100 cm and body mass index (BMI) 20 kg/m^2 (>97th percentile). On examination, vital signs, including HR, were 115/min, BP 145/75 mmHg (>95th percentile) and RR was 22/min. On physical examination, he had Cushingoid facies, buffalo hump and generalised excessive subcutaneous fat and signs of virilisation [Figure 3a and b]. There were no signs of facial acne and striae over the body.

Laboratory evaluation revealed that serum cortisol was 41.28 µg/dl (normal value 4.5–22.7 µg/dl) and serum testosterone was 219 ng/dl. Serum cortisol value after dexamethasone suppression test was 54.74 µg/dl. Serum ACTH was 1 pg/ml (normal value 10–60 pg/ml), and 24 h urinary metanephrine value was 48.33 µg/24 h (<350 µg/24 h). Serum electrolyte values were Na–143 mEq/l, K–2.3 mEq/l and Cl–100 mEq/l. Complete haemogram suggestive of Hb – 11.6 g/dl, total leukocyte count 7.17 thousand/µL and platelet count 192



Figure 1: Preoperative clinical picture showing cushingoid facies (a) and postoperatively follow-up clinical picture showing improved linear growth and normalisation of weight (b)

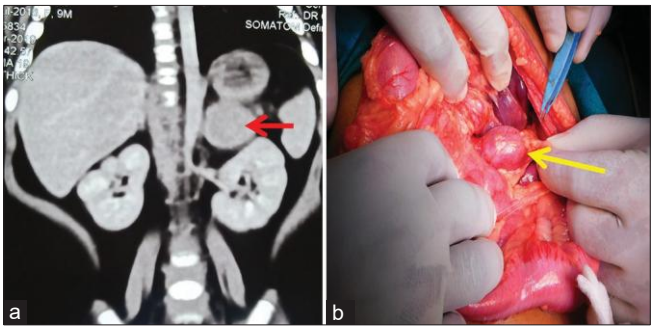


Figure 2: Contrast-enhanced computed tomography showed that a well-defined soft tissue density mass measuring 2.2 cm × 2.4 cm × 2.1 cm at the left suprarenal region (red arrow) (a) intraoperative picture of the same patient showing well-encapsulated mass arising from the left adrenal (yellow arrow) (b)



Figure 3: Preoperative clinical picture showing cushingoid facies (a) and sign of virilisation (b)

thousand/μL. Two-dimensional echo was suggestive of mild concentric left ventricular hypertrophy. Abdominal ultrasound showed a well-defined solid hypoechoic mass of size 3 cm × 3.4 cm seen in the right suprarenal region. Abdominal CECT showed that a well-defined oval heterogeneous soft-tissue lesion of size 2.7 cm × 2.3 cm × 3.4 cm was seen at the right suprarenal region [Figure 4a]. It was showing intense post-contrast enhancement. The mass abutting the liver superiorly and laterally, right crush of the diaphragm medially and the kidney inferiorly with preserved intervening fat plane. Preoperatively, the patient was managed conservatively to control BP and correct dyselectrolytemia. After hemodynamic stabilisation patient was planned for exploratory laparotomy. The child underwent exploratory laparotomy with right subcostal incision, and right adrenalectomy was performed. Intraoperative findings revealed approximately 3.0 cm × 4 cm in size well-encapsulated mass arising from the right adrenal [Figure 4b]. The mass was abutting superiorly with the liver and medially with inferior vena cava with preserved fat plane. The liver, spleen and right kidney appeared grossly normal. The tumour was resected completely without breach of the capsule. Gross pathological examination revealed a well-encapsulated globular greyish mass of size 3 cm × 3 cm × 2.8 cm and weight 15 g. Microscopic examination revealed that tumour cells arranged in organoid nests, trabeculae and focally in sheets. Tumour cells were monotonous with round, regular nucleus, compact eosinophilic cytoplasm, vesicular chromatin and inconspicuous nucleoli. There was no atypical mitosis or increased mitotic activity seen. There is no microscopic evidence of capsular or vascular invasion. Histopathological features were suggestive of an adrenocortical adenoma.

After surgery, the baby was treated with stress dosing of intravenous hydrocortisone. The post-operative period was uneventful, and discharged on maintenance doses of hydrocortisone. No adjuvant treatment was given post-operatively. During the follow-up period, his linear growth velocity improved and his weight normalised.

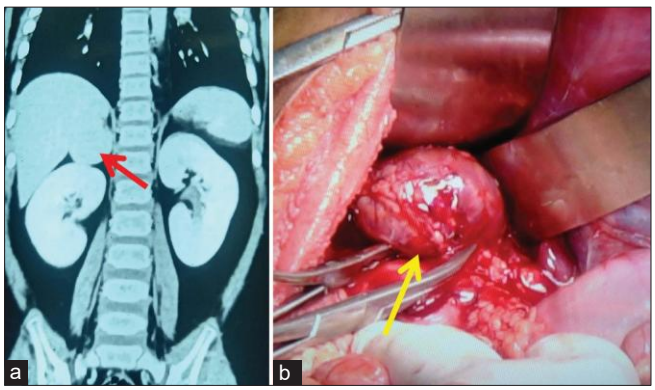


Figure 4: Contrast-enhanced computed tomography showed that a well-defined oval heterogeneous soft-tissue lesion of size 2.7 cm × 2.3 cm × 3.4 cm was seen at the right suprarenal region (red arrow) (a) and an intraoperative picture of the same patient showing well-encapsulated mass arising from right adrenal (yellow arrow) (b)

Discussion

CS in the paediatric age group is usually characterised by slowed linear growth, accelerated weight gain, facial puffiness and buffalo hump.[6] CS can be ACTH-dependent or ACTH-independent. ACTH-independent causes of CS are primary adrenocortical hyperfunction, ACTs, primary pigmented nodular adrenocortical disease, bilateral macronodular adrenal hyperplasia and iatrogenic.[7] In both cases, serum cortisol levels were elevated, whereas the ACTH level was severely suppressed, so an ACTH-independent cause of CS was suggested.

ACTs are rare in infancy, and approximately 60% of cases present between 1 and 5 years of age (60%).[8] Very few reports suggested that ACT presents in early infancy with isolated CS.[9] Both ultrasonography scan and CT scan were required for evaluation of adrenal mass but the CT scan was more sensitive. Daneman *et al*. reported that CT scan was best for evaluation as well as characterisation of an adrenal tumour.[10] Proper pre-operative preparation of the patient is essential to avoid life-threatening acute adrenal insufficiency. Due to prolonged suppression of ACTH, the contralateral adrenal gland also becomes hypoplastic. So, hydrocortisone therapy is important during the perioperative period.

In conclusion, ACT is a rare malignancy but considered as a differential diagnosis of Cushing syndrome in the paediatric age group. Although virilising features are more common, ACT may present with isolated CS. Early diagnosis, proper perioperative management, complete surgical excision of the tumour and close follow-up are crucial to improve survival.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parents have given their consent for their images and other clinical information to be reported in the journal. The parents understand that their names and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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