A CASE OF CUSHING’S SYNDROME DUE TO ADRENOCORTICAL ADENOMA WITH PUBARCHE AND OBESITY

Fatih Gürbüz, 2Atilla Çayır, 3Esra Karakuş, 4Rabia Demir, 1Fatma Demirel, 4Emrah Şenel
1Ankara Pediatric Hematology and Oncology Training and Research Hospital, Clinic of Pediatric Endocrinology, Ankara, Turkey
2Erzurum Region Training and Research Hospital, Clinic of Pediatric Endocrinology, Erzurum, Turkey
3Ankara Pediatric Hematology and Oncology Training and Research Hospital, Pathology, Ankara, Turkey
4Ankara Pediatric Hematology and Oncology Training and Research Hospital, Clinic of Pediatric Surgery, Ankara, Turkey

BACKGROUND

Adrenocortical tumors in childhood represent very rare about 0.2% of all pediatric malignancies. Cushing’s syndrome (CS) is characterized by clinical features caused by autonomous excessive glucocorticoid production from adrenal cortex. In ACTH-independent CS, the most common cause is unilateral cortisol-producing adrenocortical adenoma.

PATIENT

Fifteen months old girl was admitted with gradually gain weight, pubarche, and acne of the face since five months. There was no history of using drugs which contain steroid. Length was 74.4 cm (-1.31 SDS), weigh was 13.2 (2.16 SDS), moon facies, buffalo hump, facial plethora with acne on her forehead and cheeks. Her pubic stage was 3. In laboratory, glucose (fasting) 93 mg/dL, insulin (fasting) 24.99 μIU/mL, total testosterone 76 ng/dl, cortisol (morning) 37.69 mg/dL, ACTH (morning) 5.04 pg/ml, cortisol (evening) 30.4 μg/dL, ACTH (evening) 5.32 pg/ml, DHEA-SO4 238 pg/mL, sodium 138 mmol/L, potassium 4.53 mmol/L, WBC 20.8 10^3/mL, RBC 5.05 10^6/mL, hemoglobin 15.3 g/dL, and platelet 391 10^3/mL. After evening dexamethasone implementation, cortisol (morning) was 29.36 μg/dL. In magnetic resonance imaging identified pronounced thickening and nodularity in lateral crus of left adrenal gland.

RESULTS

Patient was diagnosed with Cushing’s syndrome due to adrenocortical tumor and administered laparoscopic left adrenalectomy. The removed mass was consistent with adrenal adenoma histologically. Postoperative fourth day control Cortisol (morning) 4.73 mg/dL, ACTH 18.7 pg/ml, and DHE-SO4 <15.0 pg/mL.

CONCLUSIONS

In adrenocortical tumors, distinguishing to adrenocortical cancer and adenoma is difficult. Herein we represent a rare case of CS because of adrenocortical adenoma with clinical features of excessive glucocorticoid production.