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THE TREATMENT OF A FUNCTIONAL ADRENOCORTİCAL CANCER WITH MITOTANE

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In the pediatric age group, the incidence of the adrenocortical cancer (ACC) is %0.2, and they are functional up to 80-90 percent. ACCs can be observed in association with Beckwith-Wideman Syndrome, congenital adrenal hyperplasia or mutations seen in tumor suppressor gene p53 as in Li-Fraumeni syndrome. The effective treatment is surgical resection, but chemotherapy and medical adrenalectomy are also options. The only choice for medical adrenalectomy is mitotane but it has negative effects on steroidogenesis.
The difficulty in the management of mitotane therapy is discussed in this case. The changes in the hormonal status of the case lead to challange in the differentiation of recurrence and adverse reactions of mitotane therapy.

Case :

The presented case, an $11^{10/12}$ years-old boy, was referred us with a 5.5*5cm solid-hypoecoic mass observed by sonography in the localization of left surrenal region. Sonography was performed because of the complaint of abdominal pain. The physical examination was all normal at presentation. Testicular volumes were 8 ml bilaterally, the penile lenght was 11cm and pubarch was Tanner grade 3. Clinical findings of cushing syndrome or over virilization were not present. In the laboratuary analysis CBC and biochemical analyses were all normal. The hormone profile was ACTH (adreno-corticotrophic hormone) <5pg/ml, 11-DOC (11-deoxyco) 11.9ng/ml, 17-OH progesteron 1.9ng/ml, cortizole 11.2 mcg/dl, DHEAS 17203ng/ml, AS 4.27 ng/ml, T.T (total testesterone) 767 ng/dl, PRA 5.1 ng/ml/hour and aldesterone 181pg/ml. Analysis of 24-hour urine specimen revealed normal catecholamine but high cortisole levels. The case was diagnosed as grade 3 ACC according to the clinic and magnetic resonance imaging (MRI) findings and treated via surgical resection. Postoperatively chemotherapy, mitotane and hydrocortisone treatments were started. In the first month of the treatment hormonal profile was all normal but TT levels started to increase (TT 650 ng/dl) by the end of second month. In the follow-up, height growth stopped and bilateral gynecomasty developed. MRI and PET (pozitron emission tomography) scans and scrotal US were negative for recurrance or metastases at six months of treatment. The clinic was diagnosed as hypergonadotropic hypogonadism (HH) due to mitotane treatment. The follow-up of the case is continuing.

Conclusion :

Mitotane treatment leads to HH via reducing the gonadal steroidogenesis. Additionally, treatment increases the levels of SHBG (sex-hormone binding globuline) and decreases the activity of 5-alfa reductase that results with high levels of testesteron but normal levels of androgene index. In our case the testesterone levels were high but on the other hand free androgen index was normal (9.9). The high levels of testesterone can be the result of metastase, recurrance or mitotane treatment adverse effect. This is a struggling problem in the management of mitotane therapy.

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