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CASE REPORT

Tuğrul Tiryaki · Zehra Aycan · Sema Hücümenoğlu
Halil Atayurt

Testis sparing surgery for steroid unresponsive testicular tumors of the congenital adrenal hyperplasia

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Abstract The association between testicular tumors/nodules and congenital adrenal hyperplasia (CAH) has been recognized for many years. Tumors are considered to be an aberrant adrenal tissue that has descended with the testes and has become hyperplastic due to ACTH stimulation. The recommended treatment consists of increasing the glucocorticoid dose to suppress ACTH secretions. If the testicular size is not reduced after suppression therapy or a side effect of glucocorticoid dose is noted, surgical intervention should be considered. We diagnosed steroid unresponsive testicular tumors of the CAH in two patients who were treated by testicular sparing tumor enucleation. We believe that testis sparing surgery is the procedure of choice for all patients with testicular adrenal rest tumor, since it maximizes future fertility potential.

Keywords Congenital adrenal hyperplasia · Testicular adrenal rest tumors

Introduction

Testicular adrenal rest tumors are increasingly reported in male patients with congenital adrenal hyperplasia (CAH) [1, 2]. Testicular masses may create diagnostic difficulties clinically and pathologically. The exact cellular origin of this tumor and the management of these patients is still controversial. When the presence of the adrenogenital syndrome is not appreciated, the mass is usually assumed to be a neoplasm and an orchidectomy is performed. The lesion may be misinterpreted as a Leydig cell tumor on pathological examination. We have presented two patients with congenital adrenal hyperplasia whose testicular tumors did not regress with suppression therapy and discussed briefly the management of CAH patients with testicular enlargement.

T. Tiryaki (✉) · H. Atayurt
Pediatric Surgery Clinic, Social Security Institution Children's Hospital, Ankara, Turkey
E-mail: httiryaki@ hotmail.com

Z. Aycan
Department of Pediatric Endocrinology, Social Security Institution Dıskapı Hospital, Ankara, Turkey

S. Hücümenoğlu
Pathology Clinic, Social Security Institution Dıskapı Hospital, Ankara, Turkey

T. Tiryaki
Atatürk Bulvarı Erciyes, Han No 109/14, Kızılay, Ankara, Turkey

Case report

Case 1

A 17 year-old boy was presented with bilateral testicular swelling of 1 month duration. A diagnosis of 11-hydroxylase deficiency had been made at the age of 15 months when he presented with dehydration, and electrolyte abnormalities. Physical examination disclosed bilateral nodular testes. Bilateral testes volume were over 25 ml and poor compliance of suppressive therapy was noted. Scrotal ultrasonography showed heterogenic testis parenchyma and multiple different sized hypoechoic nodules. On magnetic resonance imaging (MRI), a lesion filling the scrotum with dimensions of 10×8×8 cm was noted (Fig. 1). On T1a and T2a images, the mass was hypointense and was described as a rest tumor. Azospermia was found at sperm analysis. The dexamethasone dose was increased to a higher dose (1.5 mg/24 h) for replacement therapy. After 6 months, the testicular sizes were reduced but high glucocorticoid doses induced side effects. Surgical intervention was considered due to the persistence of tumor despite overtreatment. At the operation bilateral, a truffle-like noduler tumor was enucleated with testicular tissue sparing.

Histopathalogic examination of the formalin-fixed tissue showed large, polygonal, eosinophilic cells with

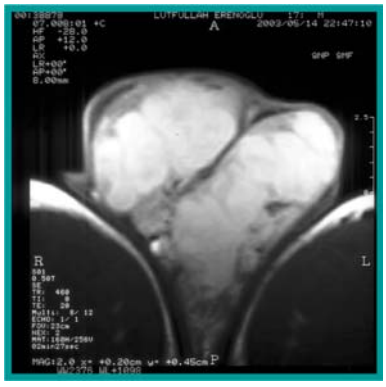

Fig. 1 Magnetic resonance imaging showed a lesion filling the scrotum with dimensions of 10×8×8 cm

round nucleus and prominent nucleolus. Reinke crystalloids and lipochrome pigment were not identified.

Case 2

A 7.1-year-old boy was presented with rapid growth, acne, and excessive hair which had been evident for 24 months. Physical examination showed a muscular, hirsute boy with a developed penis, and left testicular enlargement. Right testicular volume was 3 ml, left testicular volume was 8 ml. A diagnosis of 21-hydroxylase deficiency was established. Scrotal ultrasonography showed a 13×18 mm hypoecogenic solid mass in the middle pole of the left testes. Hypointense solid left testicular mass (10×10 mm sized) was noted on MRI. After 4 months of 30 mg/m^2/day hydrocortison suppressive medication nodules failed to regress and testis sparing surgery was performed. On gross examination the mass appeared to be a firm yellow brown nodule which localized testicular hilus with 15-mm size. Histopathologically, testicular mass cannot be distinguished from Leydig cell tumor (Fig. 2). We favored the diagnosis of Leydig cell tumor considering the response to steroid therapy. On follow-up, normal MRI and testis scintigraphical examination was noted.

Discussion

Unilateral or bilateral testicular enlargement in patients with congenital adrenal hyperplasia has been well described. Male patients with CAH (8.2%) developed testicular masses [2]. Testicular tumor of CAH is frequently bilateral, occurring syncronously. These tumors usually develop in untreated or inadequately treated boys. These tumors decrease in size with optimal steroid administration by suppression of the elevated ACTH levels [1–3].

Histopathologically, testicular masses associated with CAH resemble both adrenal cortical and Leydig cell

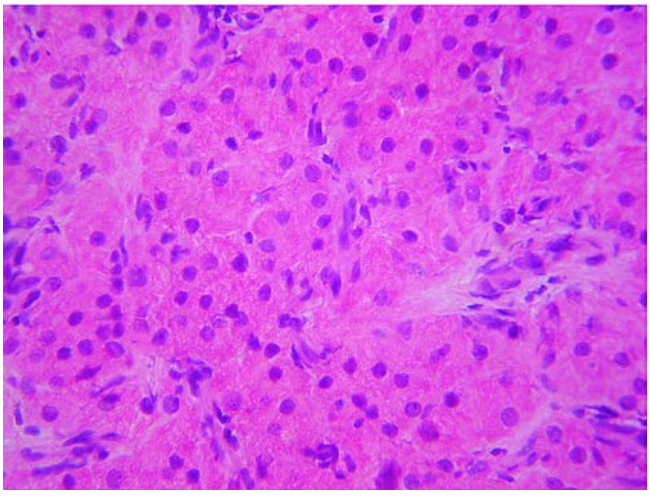

Fig. 2 Histopathologically, testicular mass did not distinguished from Leydig cell tumors

tumors. Differentiation between these two entities is very difficult. Some biochemical methods [4] have been proposed to differentiate between these two pathologies but have proved to be unreliable [5]. The diagnosis may be suspected on the basis of the history of CAH and ultrasonographic demonstration of the testicular adrenal-like tissue.

The most appropriate approach to these entities is observation of response to treatment with glucocorticoids. Regression in tumor size by suppressive treatment is suggestive to consider diagnosis of adrenal rest tissue. No changes in testicular size should be considered as true Leydig cell tumor [5, 6]. If the tumor does not shrink despite suppression treatment or if there is persistent azoospermia despite tumor reduction, surgical intervention should be considered [7].

With a history of sexual precocity in patients presenting with testicular masses, if a frozen section diagnosis of Leydig cell tumor is made at the time of biopsy, an endocrinologic evaluation is indicated to exclude unrecognized CAH to avoid an orchiectomy. Testis sparing surgery for prepubertal epidermoid cysts, teratomas, simple cysts and Leydig cell tumors has encouraging results at long-term follow-up besides psychological and cosmetic advantage for the developing child [8, 9].

The natural course of the testicular adrenal rest tumors and their impact on fertility are unknown in young patients with CAH. Our observations suggest that decrease in the size of tumor can be obtained by aiming adrenal oversuppression, but this usually requires high glucocorticoid doses and induces side effects. Because of the high prevalence of testicular tumors in these patients, we would recommend performing testicular ultrasonography in all male patients with CAH on a regular basis. And also testicular adrenal rest tumors can be the first manifestation of CAH. Enucleation of the tumor is the surgical management of steroid unresponsive testicular tumor of CAH. Testis sparing surgery is an

important consideration, since male individuals with CAH are potentially fertile.

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