

TESTICULAR ADRENAL REST TUMORS. A Case Report

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Introduction

Congenital adrenal hyperplasia caused by inherent deficiency of an adrenal enzyme, mostly due to deficiency of 21 hydroxylase, leads to defective levels of glucocorticoid synthesis. In addition, aldosterone synthesis is also effected. Subsequent increases in ACTH levels lead to adrenal cortex hyperplasia. Increased corticotropin levels prevent involution of aberrant adrenal cortical cells which migrate with gonadal tissues in fetal life. By means of uncontrolled high corticotropin levels, this ectopic tissue may develop in to adrenal rest tumors.

Case

The patient had been diagnosed with congenital adrenal hyperplasia (CAH) at age 5 years because of precocious of puberty. He had been receiving dexamethazone and fludrocortisone therapy over the course of his life. When he was 16 years-old, a scrotal ultrasound had been performed for the evaluation of bilateral testicular pain which revealed multiple sharply marginated, hypoechoic masses throughout both testes. A biopsy of left testis reported as "Leydig cell tumour". He refused orchiectomy and exited the follow-up of the prior clinic. After 4 years, he was admitted to our endocrinology clinic at the age of 20 years. A review of his story and laboratory findings showed that he had 21 hydroxylase deficiency, bilateral tumor of the testes. His compliance to glucocorticoid treatment was poor. The first biopsy of testis and a new biopsy of left testis was evaluated by the same pathologist and reported as a proliferation of polygonal cells with abundant granular, eosinophilic cytoplasm. Reinke crystals were absent, Ki67 %1-2, CD56 (+). Immunohistochemically, synaptophysin was positive, inhibin-alpha was focally positive.

CONCLUSIONS

.. Testicular lesions in the setting of CAH are commonly referred to as testicular adrenal rest tumors. In prenatal life, the adrenals develop in the immediate vicinity of the gonads, and separation of both does not evolve until the adrenal groove becomes prominent. Before that moment, adrenal cortical tissue may adhere to the gonad. This aberrant adrenal tissue may then descend with the testis or ovary along the course of their supplying arteries. Therefore, it is possible that the testicular tumors of CAH originate from aberrant adrenal tissue. Adrenal rest tumours can be the first manifestation of CAH, as Rutgers et al reported undiagnosed CAH in 18% of adrenal rest tumour cases. They are frequently benign, multiple and bilateral. The tumors are most prevalent in younger adult males with a peak incidence between 20-40 years of age. First line imaging modality in the detection and surveillance of these tumors is ultrasonography. Histologically, testicular tumors of the adrenogenital syndrome are commonly mistaken for Leydig cell tumors as in our case. Clinical follow-up without orchiectomies is recommended unless untreated nodules of adrenal rests expand and destroy the testicular parenchyma, resulting in low testosterone production and infertility. Although poor fertility in male CAH patients was reported, untreated male CAH patients were reported to be normally fertile. Patient compliance to treatment is important since the presence of adrenal rest tumors are suggestive of suboptimal hormone replacement therapy. When the tumor is unresponsive to steroid therapy, surgical treatment should be considered, preferably by a testis-sparing procedure, instead of orchiectomy. Also, cryopreservation of the semen can be offered, because fertility prognosis is yet uncertain.

References

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