Doppler ultrasonography and magnetic resonance imaging findings of testicular adrenal rest tissue in a patient with 11 β hydroxylase deficiency. Case report.

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Abstract

Benign intratesticular lesions are rare; thus, radiological diagnosis is important to avoid unnecessary surgical intervention. Testicular adrenal rest tumors (TART) are the nodular testicular lesions deriving from the adrenal remnant tissue reported in patients with congenital adrenal hyperplasia. We present ultrasonographic (US) and magnetic resonance imaging (MRI) findings of intratesticular adrenal rests in a 16-year-old patient with 11 β hydroxylase deficiency. Scrotal ultrasonography demonstrated bilateral heterogenous hypoechoic lesions located around mediastinum testis and highly vascularized on power Doppler US. On MRI the lesions were hyperintense on T1-weighted and hypointense on T2-weighted images and enhanced brightly on post-contrast images.

Keywords: Doppler ultrasonography, MRI, Testicular Adrenal Rest Tissue, 11 β hydroxylase deficiency

Introduction

Congenital adrenal hyperplasia (CAH) describes a group of inherited autosomal recessive disorders characterized by enzyme defects in the steroidogenic pathways. Therefore that leads to decreased biosynthesis of steroid hormone such as cortisol, aldosterone, and androgens [1]. CYP21 (21-hydroxylase) deficiency is the most common cause of CAH [2]. Steroid 11β hydroxylase deficiency accounting for less than 5 % of cases and is the second most common cause of CAH. It is characterized by the over production of adrenal androgens and deoxycorticosterone which lead to virilization of female fetuses, pseudoprecocious puberty in male infants, and hypertension in both genders [3]. Increased adrenocorticotropic hormone (ACTH) synthesis results in hyperplasia of ACTH-sensitive tissues in adrenal glands and other sites such as the testes. Aberrant adrenal tissue that has become hyperplastic because of elevated ACTH cause testicular masses known as testicular adrenal rest tumors (TARTs) [4,5]. Herein we present the case of a 16-year-old patient with CAH due to 11 β-hydroxylase deficiency who presented with TARTs.

Case report

Sixteen-year-old male patient followed up by out center clinic for congenital adrenal hyperplasia was admitted the urology clinic presenting bilateral testicular pain a lump at the superior part of scrotum, and excessive pubescence. On his physical examination bilateral mass lesion at the superior part of testis and bilateral testicular pain with palpation was noticed. His blood pressure was normal. Renin and aldosterone values were normal. At ultrasonographic (US) examination in both testis heterogeneous hypoechoic lesions extending by rete testis to parenchyma, with irregular borders, internal local hyperechoic appearances, and hypervascularization were found (fig 1). On MRI isointense signal changes on T1 weighted images and hypointense signal changes on T2 weighted images at bilateral mediastinum of the testis were described. Brightly contrast enhancement was seen on these lesions (fig 2). There was also dilatation at pam-
piniformis veins showing varicocele. The remaining testis tissue was normal. The bilateral testicular lesions were interpreted as testicular adrenal rest tumor. After steroid treatment control US examination was performed showing slightly decrease in size and prominent decrease in vascularization (fig 3).

**Discussions**

TARTs are one of the known complications of CAH. Their reported prevalence is up to 94% of CAH adults, and they may already appear during childhood. Embryological development of the adrenal cortex occurs in close proximity to the gonads. It has been suggested that adrenal rest tumors consist of adrenal tissues localized in the scrotum, within the testicles.

In patients with CAH, TARTs are usually seen bilaterally. With adequate steroid replacement therapy, tumor size decreases [6]. Due to the location of the tumors within the rete testis, TARTs are difficult to palpate. Therefore, usually only tumors with a size of more than 2 cm are detectable by palpation. US and MRI are equally good methods for detection and characterization of the tumors. On US, they are characteristically located around the mediastinum testis and do not deform the shape of the testis. The lesions are mostly bilateral and hypoechoic compared with the normal parenchyma. Depending on the extent of fibrotic changes, an attenuation of the sound may be seen [7]. Few publications have reported MRI findings in testicular adrenal rests. Like adrenal cortex, testicular adrenal rests are isointense with muscle on T1- and T2-weighted images. In relation to the testicular parenchyma, the lesions appear either iso- or hyperintense on T1-weighted and hypointense on T2-weighted images, without a capsule or a pseudocapsule. Homogenous enhancement occurs after the injection of gadolinium [7]. Until now, intensive glucocorticoid treatment is the optional treatment in patients with TART. Intensive glucocorticoid therapy by suppressing ACTH secretion may lead to reduction of the tumor size, thereby improving testicular function [8].

Although benign intratesticular lesions including TARTs are rare, their imaging findings are very similar...
to malignant testicular lesion, so differentiation between them is very important to avoid unnecessary surgical intervention [9].

Our case suggests that radiological evaluation of testes, even in the presence of normal physical examination findings, should be included in periodical follow-up of patients with congenital adrenal hyperplasia.

References