CASE REPORT

Bilateral testicular adrenal rest tissue in congenital adrenal hyperplasia: US and MR features

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ABSTRACT. We describe magnetic resonance (MR) and ultrasonography (US) features of bilateral testicular adrenal rest tissue in a 20-yr-old man with congenital adrenal hyperplasia (CAH). Scrotal ultrasonology detected bilateral homogeneous hypoechoic lesions with well-defined margins and without evidence of sound attenuation. MR is useful in defining the size of lesions, because the contrast resolution is better than with sonography and allows an accurate definition of the extent of disease. This case suggests that US evaluation should be included in periodical follow-up of patients with CAH, while MR may be used in the case of rapid increase in the size of the testicular mass.
(J. Endocrinol. Invest. *24:* 529-531, 2001)

INTRODUCTION

It is well known that testicular "tumors" (testicular masses representing adrenal rest tissue) may be sonographically identified in patients with congenital adrenal hyperplasia (CAH) or in other conditions associated with a high level of circulating ACTH, such as Addison's disease and Cushing's syndrome (1).
The true frequency of testicular masses in patients with CAH is unknown because they are not typically screened by testicular sonography; in fact, while in 1963 it was reported that testicular adrenal rest was present in all patients with CAH (2), in a more recent study a prevalence of 29% is reported (3, 4) and for others it is a rare clinical condition described in approximately 50 cases (5).
This report aims to describe imaging features [ultrasonography (US) and magnetic resonance (MR)] of one patient affected by adrenogenital syndrome and bilateral intratesticular adrenal rest.

Key-words: Testicular adrenal rest, congenital adrenal hyperplasia.
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Accepted September 18, 2000.

CASE REPORT

A 20-yr-old man was admitted to our Endocrine Unit for poorly controlled 21-hydroxylase deficiency [ACTH 720 ng/l, 17-hydroxy-progesterone (17-OHP)>20 ng/ml]. He had CAH (classic form with salt loss) diagnosed early in the neonatal period, after presenting hypoglycemia, shock and chemical findings compatible with adrenal insufficiency. The patient was in therapy with dexamethasone (0.25 mg/*die*) and fludrocortisone (0.1 mg/*die*). Testes were normal on palpation.
Scrotal US, both standard gray scale and color Doppler, was performed by using 7.5 MHz linear-array transducer with commercially available sonographic equipment (model AU590 A unit, Ansaldo Medical System, Genova, Italy).
US detected bilateral testicular lesions, homogeneously hypoechoic, of oblong shape, with well defined margins and without sound attenuation. Both lesions were peripherally located, close to the mediastinum testis, surrounded by normal testicular parenchima and without alteration of the profile.
Color Doppler evaluation showed little vascularity in the peripheral portion of the lesions (Fig. 1A and B).
MR was performed with a 1 tesla Siemens and an anular surface coil by using coronal and transverse plane with T1- and T2-weighted images and contrast-enhanced T1-weighted images. The lesions were isointense in the T1-weighted images; hypointense in the T2 images and showed marked enhancement in T1 images after contrast media (Magnevist-Schering) (Fig. 2A, B and C).

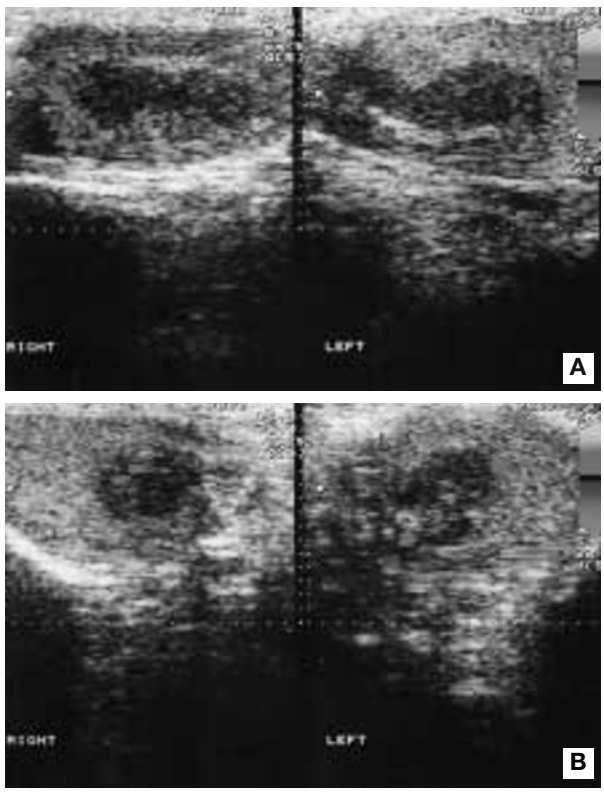

Fig. 1 - *Longitudinal (A) and axial (B) sonograms of the right and left testes show bilateral hypoechoic lesion close to the mediastinum testis with poor vascularity in the peripheral portions of the lesions.*

A fine needle biopsy of the testicular nodules was performed. The small fragments were fixed in buffered formalin and routinely embedded in paraffin. Microscopically, the sections showed clusters of cells with abundant eosinophilic cytoplasm without Reinke crystals. The absence of lipochrome pigment and Reinke's crystals suggests a diagnosis of testicular tumors of adrenogenital syndrome.

DISCUSSION

A sonographic diagnosis of testicular adrenal rest can be suggested in conditions associated with high level of ACTH, such as CAH, Addison's disease and Cushing's syndrome

However, these lesions do not show a specific sonographic pattern, because single and unilateral or multiple and bilateral hypoechoic lesions may represent seminomatous and non-seminomatous tumors, lymphoma, leukemia and other testicular lesions.

Testicular adrenal rests are in most cases hypoechoic lesions, peripherally located close to the mediastinum testis, of oblong shape, associated with controlateral lesion, causing no alteration of the testicular profile and not clinically palpable when small (6, 7).

The hypoechoic pattern is present in all such lesions as reported in the literature. Various degrees of acoustic shadowing due to the extensive fibrosis of these lesions may occur (6, 8, 9), but we did not find any.

Most of the lesions are located inside or close to the testicular hilus (86%) and they are normally bilateral (83-100% of the cases) (6-9).

The lesions may be palpable (reported in two-thirds of the lesions) or unpalpable, under 2 cm in diameter (8), and in spite of their peripheral location do not cause alteration of the testicular profile.

At color Doppler the lesions may be iso/hyper/hypovascular relative to the normal testis, without correlation between diameters or volume of the lesion and vascularity (8, 9). However, all testicular masses contained vascular structures coming from the normal testis, without a change of its course or caliber (8, 9).

MR is as useful as sonography in the detection of testicular adrenal rest tissues, but not in their differentiation from malignant seminomatous tumors (8).

In our case MR helped obtain a precise definition of the size of the lesions, because it has a better contrast resolution than sonography and because it allows an accurate definition of the extent

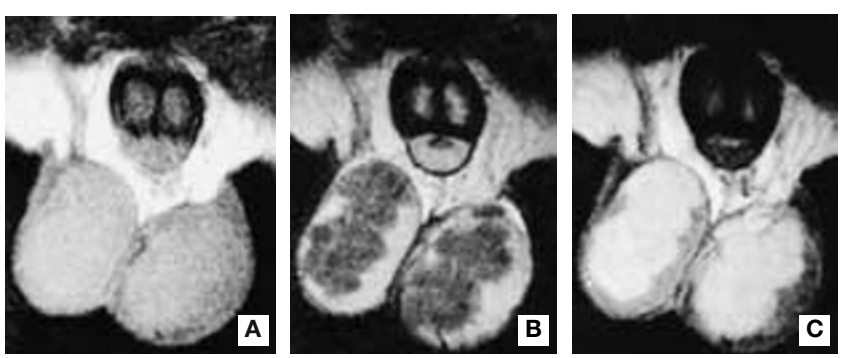

Fig. 2 - *Magnetic resonance shows: A) unenhanced coronal T1-weighted spin-echo with no evidence of the testicular lesions; B) in T2-weighted images lesions are marked hypointense compared to the normal testis; C) in enhanced T1-weighted images after paramagnetic contrast media infusion lesions show marked and diffused hyperintensity.*

of the disease. It is also less operator-dependent than sonography.
However, MR should be used only in selected cases, because it is a very expensive and time consuming way to follow-up patients.
Pathological differentiation between testicular adrenal rests and Leydig cell tumors is possible only in presence of Rainke's crystalloids, (which are also found in 40% of Leydig cell tumors) otherwise they are pathologically indistinguishable (7, 8).
Although a variety of origins have been suggested for these lesions, they most likely arise from the hilar pluripotential cells, which proliferate as a result of the elevated level of ACTH (8) or may derive from particular ACTH-dependent Leydig cells or from staminal cells with potentiality to differentiate into either adrenal-like or Leydig-like cells, depending upon the stimulated tropin (ACTH or LH) (10-12).
Therefore, we believe that testicular tumors are not autonomous neoplasms but may be dependent on the elevated levels of ACTH. The tissue will hypertrophy when there is inadequate glicocorticoid replacement and it may atrophy under the influence of a high dose of glicocorticoids. According to some authors, however, there is no relationship between clinical control (based on the 17-OHP level) at the moment of the sonographic examination and changes in the size of the testicular adrenal rest tissue (4). Its increase, therefore, suggests the need for more aggressive treatment.
Malignant degeneration is very rare but testicular biopsy needs to be performed when the lesion's size is shown to increase, on clinical and sonographical follow-up. In this case, the first-line therapy should be a higher dose of glicocorticoids with sonography repeated after one month to evaluate the response (4).
All patients with CAH should be submitted for sonographic evaluation, especially between 1st and 2nd decades of life since most testicular tumors are detected in this period. Once a testicular mass is sonographically diagnosed it must be followed up (4).
All patients with bilateral testicular tumors should undergo endocrinological evaluation to exclude the possibility of CAH, even in the absence of clinical symptoms.

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