Testicular adrenal rest tissue in two brothers with congenital adrenal hyperplasia

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Section: Uroradiology & genital male imaging
Area of Interest: Genital / Reproductive system male
Procedure: Diagnostic procedure
Imaging Technique: Ultrasound
Special Focus: Tissue characterisation Case Type: Clinical Cases
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Patient: 23 years, male

Clinical History:

The patients, 23 and 28 years old, with classic congenital adrenal hyperplasia diagnosed and treated since birth, were referred by their endocrinologist to rule out testicular adrenal rest tissue.

Imaging Findings:

In both patients testicular ultrasound revealed bilateral, lobulated, hypoechoic tumour-like masses located in the periphery, with their long axis disposed parallel to the long axis of the testes. Posterior acoustic shadow was noted, probably due to fibrotic changes. Colour Doppler ultrasound showed increased vascularity, with low resistant flow vessels.

Discussion:

Congenital adrenal hyperplasia is an autosomic recessive disorder caused by a deficiency of one of five enzymes required for the synthesis of cortisol in the adrenal cortex. In more than 90 percent of cases the deficiency is steroid 21-hydroxylase [1, 2].

Patients with this disorder have low levels of both glucocorticoids and mineralocorticoids, which leads to excess adrenocorticotropic (ACTH) secretion by the pituitary gland, with consequent adrenal hyperplasia. Testicular adrenal rest tissue (TART) is a benign lesion seen as an intratesticular mass in patients with CAH. Adrenal rests arise from aberrant adrenocortical cells that migrate with gonadal tissues in the fetus [3], although it may be seen at several sites throughout the body [4].

Imaging plays an important role in the detection and surveillance of TART. Ultrasound (US) is the modality of choice in the detection, because it is as sensible as MR, widely available and less expensive [2, 4] MR may be superior in differentiating between multiple small, discrete lesions and a larger, solitary multilobar lesion and is often preferred when the precise extent of disease is required [2].

In US, they are in most cases multifocal hypoechoic lesions, peripherally located close to the mediastinum of the testes, and usually bilateral [5, 6]. Larger lesions may have central regions of increased echogenicity. Occasionally,
posterior acoustic shadow has been described due to fibrosis [6, 7]. They may vary in size, ranging from 4 to 38 mm [2]. These lesions usually are hypovascular or normovascular, but may be infrequently hypervascular (2). A “spoke-wheel” pattern of increased vascularity has been reported [6, 7].

Imaging findings may overlap with primary germ cell or gonadal tumours, but they are generally unilateral [2]. TART can be indistinguishable from Leydig cell tumour (LCT), which is bilateral only in 3% of cases and associated with distinct endocrine abnormalities and immunohistochemical features, while TART typically appears in association with CAH and is bilateral in more than 80% of cases [8].

If the diagnosis cannot be established by imaging, testicular vein sampling may show elevated cortisol levels compared with peripheral blood levels [2]. Biopsy can be made in a surgically exposed testis [3].

TART may regress with administration of exogenous glucocorticoids in doses high enough to suppress ACTH [2, 4], and can be monitored with US and measuring 17-hydroxyprogesterone and androstenedione levels. Untreated TART may compress and distort normal background testicular parenchyma, and can result in infertility [2].

Differential Diagnosis List: Testicular adrenal rest tumour in patients with congenital adrenal hyperplasia., Leydig cell tumour, Synchronous malignant testicular tumours

Final Diagnosis: Testicular adrenal rest tumour in patients with congenital adrenal hyperplasia.

References:

Description: 23-year-old man: Testicular ultrasound shows polylobulated, sharply marginated, hypoechoic bilateral, tumour-like lesions, located in the periphery of the testes, disposed parallel to their long axis, with increased sound absorption that suggests fibrotic changes. Origin: Department of Radiology, Morales Meseguer Hospital, Murcia, Spain
Description: Colour Doppler ultrasound of the previous patient manifests increased vascularity within the lesion. Origin: Department of Radiology, Morales Meseguer Hospital, Murcia, Spain
Figure 3

**Description:** 28-year-old man: Testicular ultrasound shows bilateral polylobulated hypoechoic tumour-like lesions, located in the periphery and oriented parallel to the long axis of the testes. **Origin:** Department of Radiology, Morales Meseguer Hospital, Murcia, Spain