Case 17590

Testicular adrenal rest tumors
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Section: Uroradiology & genital male imaging
Area of Interest: Genital / Reproductive system male
Paediatric
Imaging Technique: Ultrasound
Imaging Technique: Ultrasound-Colour Doppler
Case Type: Clinical Cases
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Patient: 12 years, male

Clinical History:
A 12-year-old male patient with the neonatal diagnosis of congenital adrenal hyperplasia presented to our institution with a 6-hour history of acute scrotal pain after a minor blunt trauma during rugby practice. The patient denied nausea or vomiting. Physical examination was normal.

Imaging Findings:
Scrotal ultrasound assessment was performed and no signs of testicular or epididymis fracture, testicular or scrotal wall hematoma were present. Signs of haematocele were also absent.

Ultrasound revealed one round and homogeneously hypoechoic nodule in each testicle. Both nodules, measuring 6 mm and 4 mm, were adjacent to the mediastinum testis. They were hypervascular on colour Doppler imaging.

Discussion:
Serum testicular tumor markers (alpha-fetoprotein, beta human chorionic gonadotropin and lactate dehydrogenase) were not elevated. Given the known history of congenital adrenal hyperplasia (CAH), the bilateral findings and the lesions characteristics and location, the diagnosis of testicular adrenal rest tumors (TARTs) was considered likely.

CAH refers to a group of autosomal recessive disorders in steroid and glucocorticoid synthesis secondary to an enzymatic deficiency. In over 90% cases, the deficient enzyme is 21-hydroxylase, while in other cases the deficient enzyme is 11-hydroxylase [1]. The impaired production of cortisol and aldosterone leads to overproduction of adrenocorticotropic hormone (ACTH), adrenal hyperplasia, and excess adrenal androgen production [2].

Male patients with CAH may develop TARTs, with an increasing prevalence rate observed during and after puberty [1]. The etiology of these benign lesions is unclear, but possibilities include adrenal rests and pluripotent ectopic adrenal cortex remnant cells of the testicular stroma which differentiate and grow under the influence of chronically elevated ACTH [2].

TARTs are usually asymptomatic but can grow in size, compress and replace normal testicular parenchyma, leading to disorders in spermatogenesis and infertility [2].
Ultrasoundography is considered to be the first-line imaging modality for detection, characterization and follow-up of TARTs [2]. Ultrasound imaging features are multiple, bilateral and well-marginated masses located along the mediastinum testis. These masses are more frequently hypoechoic, but they can be heterogeneously hyperechoic with shadowing [3]. Most of the lesions have positive colour flow on Doppler imaging [4]. Some of these ultrasound features can overlap with other testicular lesions. The main differential diagnostic considerations for TARTs are malignancy (both primary and metastatic disease), infection, and sarcoidosis [2]. The great majority of patients with TARTs will have bilateral testicular involvement, whereas other similar appearing entities including Leydig cell tumours will almost always have unilateral involvement [2, 5].

Exogenous glucocorticoids are used for CAH and TART treatment by suppressing ACTH production [1]. Shrinkage or disappearance of TARTs can be seen after steroid therapy [1].

The recognition of this benign entity in the adequate clinical context will help prevent unnecessary orchiectomies.

In our case, one year after the diagnosis, the patient is asymptomatic and the size and number of lesions remain stable at follow-up ultrasound.

Written informed patient consent for publication has been obtained.

**Differential Diagnosis List:** Testicular adrenal rest tumors, Sarcoidosis, Leydig cell tumours, Metastatic disease

**Final Diagnosis:** Testicular adrenal rest tumors

**References:**


Figure 1

Description: Ultrasound images of the right (A) and left (B) testicles show homogeneously hypoechoic and well-circumscribed similar nodules. Origin: Department of Radiology, Hospital Dona Estefânia, Centro Hospitalar e Universitário de Lisboa Ocidental, Portugal, 2021
Description: Ultrasound images of the right (A) and left (B) testicles show homogeneously hypoechoic and well-circumscribed similar nodules. 

Origin: Department of Radiology, Hospital Dona Estefânia, Centro Hospitalar e Universitário de Lisboa Ocidental, Portugal, 2021
Figure 2

Description: Color Doppler ultrasound images of the same nodules in the right (A) and left (B) testicles show intrallesional color flow. 

Origin: Department of Radiology, Hospital Dona Estefânia, Centro Hospitalar e Universitário de Lisboa Ocidental, Portugal, 2021.
Description: Color Doppler ultrasound images of the same nodules in the right (A) and left (B) testicles show intrallesional color flow. 

Origin: Department of Radiology, Hospital Dona Estefânia, Centro Hospitalar e Universitário de Lisboa Ocidental, Portugal, 2021