Adrenal Nodular Hyperplasia with so Called Testicular Tumor of Adrenogenital Syndrome (Adrenal Rests of Both Testes): a case report and review of literature

YU-HSIU JUAN  YING-CHUN CHIU  GUO-SHU HUANG  CHIN-JIUN Wu

Department of Radiology, Tri-Service General Hospital

Testicular tumor of adrenogenital syndrome, also being called adrenal rests, is rare bilateral intra-testicular tumors highly associated with congenital adrenal hyperplasia (CAH). Due to the benign nature of testicular adrenal rests and effective response to steroid replacement therapy, unnecessary testicular biopsy and orchiectomy should be avoided in these patients. We present a case with bilateral intratesticular masses and incidental adrenal nodular hyperplasia.

CASE REPORT

A 22-year-old male presented with palpable painless hardness mass lesions over bilateral testes for two weeks. He had no history of trauma or other systemic disease. General physical examination was normal. Laboratory studies, including complete blood count, routine urinalysis, blood coagulation studies, blood biochemistry tests, carcinoembryonic antigen (CEA), and α-fetoprotein (AFP) tests were normal. Endocrine evaluation revealed normal thyroid function tests and values of testosterone, estradiol, follicle-stimulating hormone, luteinizing hormone, cortisol, and prolactin were normal in limits. The ACTH level (61.3 pg/mL; normal range: 9.0~52.0 pg/mL) was found to be elevated. The semen analysis showed azoosperma. Chromosomal analysis was normal as 46,XY. The scrotal ultrasonography (US) revealed normal size of both testes and several round to oval hypoechoic lesions, 8 mm ~ 1.7 cm in diameter, in bilateral mediastinum testes. They were sharply delineated from the normal parenchyma and had no acoustic attenuation or enhancement (Fig. 1a, 1b). Power
Figure 1. a, b. Longitudinal grey-scale sonogram of right (a) and left (b) testes shows several discrete hypoechoic nodules (arrows) in the mediastinum testis. c, d. Color flow Doppler sonogram reveals mild vascularity within these lesions.

Figure 2. a. Coronal T1-weighted (TR/TE/excitation, 736/12/1) spin-echo MR image shows isointense or hypointense nodules relative to normal testicular parenchyma. b. Coronal T2-weighted (3000/96/1) MR image these lesions (arrows) stand out as hypointense nodules against bright testicular parenchyma. c. Coronal post-gadolinium T1-weighted (736/12/1) MR image reveals marked enhancement of the lesions (arrows).
Doppler US showed high vascularity within the lesions (Fig. 1c, 1d). On magnetic resonance (MR) imaging, the lesions were isointense or hypointense on T1-weighted (Fig. 2a) and hypointense on T2-weighted MR images (Fig. 2b) as compared with normal testicular parenchyma. After injection of the gadolinium, lesions showed marked and homogeneous enhancement (Fig. 2c). An incidental hypoechoic nodular lesion in the right suprarenal region was found in whole abdominal US (Fig. 3). Subsequent whole abdominal computed tomography (CT) study (Fig. 4a, 4b) and MR scan (Fig. 5a-5e) revealed that this lesion located at right adrenal gland and the imaging features were not characteristic for adrenal adenoma. Due to uncertainty of the nature of the adrenal tumor and testicular masses, right adrenalectomy and bilateral testicular biopsy were carried out by urologists. Histopathologic examination showed nodular hyperplasia of the right adrenal gland and nests of ectopic adrenal cortical cells within bilateral testicular masses. The final diagnosis was adrenal nodular hyperplasia with bilateral intratesticular adrenal rests.

**DISCUSSION**

TTAGS often present as bilateral testicular masses or increased testicular size associated with elevated ACTH. This lesion most commonly reported in patients with CAH, occasionally with Addison’s disease and Cushing’s syndrome [4]. A review of Rutgers et al documented that 18% of patients with adrenal rest tumors presented with no previous diagnosis of CAH [8]. Pathologically, testicular adrenal rests are hyperplastic adrenal cortical tissue originating from aberrant adrenal tissue that adheres to the gonads and descends with the testes during the developmental stages [2]. Presentation occurs in adolescence or early adulthood. The recommended

![Figure 3. The grey-scale abdominal sonogram shows an incidental homogeneous hypoechoic nodule (arrow) over the right suprarenal fossa.](image)

![Figure 4. a. Axial non-contrast enhanced CT (NECT) image shows an isodense nodule (arrow) (Hounsfield unit H.U.: 23) of the right adrenal gland. b. Axial contrast enhanced CT (CECT) image reveals homogeneous enhancement of this nodule (arrow) (H.U.: 67).](image)
treatment consists of increasing the glucocorticoid dose to suppress ACTH secretions. If the testicular size is not reduced after suppression therapy, surgical intervention should be considered [5].

Correct identification of testicular adrenal rests is important to avoid unnecessary invasive procedures such as biopsy and orchietomy. Unfortunately, there is no reliable imaging method for the diagnosis. Even histologically, adrenal rest tissue cannot always be distinguished from Leydig cell tumor [6]. On imaging studies, the scrotal US showed bilateral variable-sized hypoechoic nodules in the mediastinum testis with various vascularity. The reported MR features of testicular adrenal rests included isointense to normal testicular tissue on T1-weighted images and hypointense on T2-weighted images. The lesions are usually isointense to muscle on T1-weighted images and T2-weighted images, resembling normal adrenal glands. Homogeneous enhancement occurred after injection of gadolinium [7]. These imaging features may be helpful for differentiation of adrenal rests from other testicular tumors but not pathognomonic.

The most common reported bilateral intratesticular tumor is lymphoma, which occurs in a much older population and is often accompanied with systemic symptoms such as weight loss, anorexia, fever, and weakness. Other malignant tumors involving bilateral testes include leukemia, metastases, Leydig cell tumor, and germ cell tumors, which have been documented with low incidence [1]. Benign lesion such as granulomatous orchitis may also have similar

**Figure 5.** (a, b) Coronal T1-weighted (130/4.1/1) (a) and T2-weighted (4.4/90/1) (b) MR images show a right adrenal tumor (arrow) with isointensity relative to muscle. c. Coronal post-gadolinium T1-weighted (130/4.1/1) MR image reveals homogeneous enhancement of this lesion (arrow). (d, e) Axial T1-weighted MR in phase (176/5.3/1) (d) and opposed phase (176/2.7/1) (e) gradient echo sequence demonstrates no significant signal “drop-out” of this adrenal lesion (arrow), not fitting lipid-rich adenoma.
imaging presentation [1, 2]. Hence, patient history and clinical presentation are important for making diagnosis of bilateral testicular masses.

In summary, we presented a young man of bilateral intratesticular tumors with adrenal nodular hyperplasia. The diagnosis of TTAGS should be kept in mind to prevent unneeded intervention procedures.

REFERENCES

腎上腺結節樣增生合併兩側睾丸的腎上腺遺跡：
病例報告及文獻回顧

阮郁修  邱盈峻  黃國書  吳清俊

三軍總醫院  放射診斷部

睾丸的腎上腺遺跡是很罕見的雙側睾丸腫瘤，而且和先天性腎上腺增生有高度的相關性。由於腎上腺遺跡是個良性的疾病同時對於類固醇的治療有相當不錯的反應，所以我們應該避免這類的病人接受不必要的睾丸切片甚至是睾丸切除。我們在此介紹一位年輕男性有著雙側睾丸腫瘤和意外發現的腎上腺結節樣增生。