Testicular Adrenal Rests Tumors: Imaging Appearance and Differential Diagnosis

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Learning objectives

The purpose of this educational exhibit is to:

1- Review the embryological development of the male gonad and the adrenal gland;
2- Discuss the most common findings at US and MR imaging that can help to establish an accurate diagnosis;
3- Report our experience with testicular adrenal rests tumors in patients with congenital adrenal hyperplasia;
4- Discuss the differential diagnosis of a bilateral testicular lesion.

Background

Embriology

The adrenal glands and the gonads share a common embryological origin.

During the 5th and 6th week of the embryological development, mesodermal cells from the posterior abdominal wall, located between the root of the mesentery and the urogenital ridge, proliferate and give rise to the adrenal cortex primordium. By the 7th week of embryological development, neural crest cells from the sympathetic ganglion migrate towards the center of the adrenal cortex primordium to form the adrenal medulla.

The gonads originated from the urogenital ridge. At the beginning of the 5th week of the embryological development, primordial germ cell migrate from the hindgut to the medial aspect of the urogenital ridge. The urogenital ridge then enlongate caudally and growth in size to give rise to the gonads.

Diverse transcriptional factors, endocrine markers and signaling pathways are required for the individualization and the differentiation of the adrenal glands and testis. Although the etiology is not completely understood, during this interrelated embryological developmental period, some cells destined to become adrenocortical cells may nestle within the descending gonad.
Testicular adrenal rests tumors (TART) are benign lesions that develop due to overstimulation of the ectopic adrenal remnants within the testis.

They are commonly found in individuals with congenital adrenal hyperplasia (CAH). Although the incidence of testicular adrenal rests varies in CAH, it usually increase with age (might be found in up to 94% of the adults with CAH).

Testicular adrenal rests can be normally found in the testis and surrounding tissues in 5% to 15% of the newborns. In normal individuals, the testicular adrenal remnants become atrophic during development and are found in less than 1% of the normal adults.

**Congenital Adrenal Hyperplasi (CAH)**

CAH is a group of inheritable metabolic diseases that affect the adrenocortical steroid synthesis. In most cases, it is related to a defect of the *CYP21A2* gene leading to 21-hydroxylase deficiency. The result is insufficient production of cortisol by the adrenal cortex and, in some cases, also inadequate aldosterone synthesis.

Nowadays, the CAH is diagnosed at an early age soon after birth and the appropriate treatment has significantly reduced the morbidity and mortality. However, the lack of the cortisol' feedback to the hypothalamus nucleus and pituitary gland results in persistently elevated serum levels of the adrenocorticotropic hormone (ACTH). The chronically elevated ACTH levels stimulates the adrenal remnants in the testis to become hyperplastic.

Individuals with Addison's disease, Cushing's and Nelson's syndromes might also present TART.

**Findings and procedure details**

**Testicular Adrenal Rests Tumors**

Imaging plays an important role in the detection and surveillance of testicular adrenal rest tumors. Ultrasound (US) is generally considered the first imaging modality for evaluation of TART.
Ultrasound

Ultrasound usually demonstrated hypoechogenic, intratesticular masses, usually with ill-defined borders and in most cases bilateral. Typically these masses are located near the mediastinum testis. (Fig 1)

Testicular adrenal rests tumor varies in size, from millimeters to a few centimeters (commonly 3 to 40 mm), however they usually don't cause architectural distortion of the surrounding testicular parenchyma or deformation of the testicular morphology. (Fig 2)

Fig. 1: Testicular adrenal rests tumors. 3 months old male with CAH. Sagittal US images demonstrated a small nodular hypoechogenic intratesticular lesion, near the mediastinum testis, that present increased vascularity on color-Doppler sonographic evaluation compared to the adjacent testicular parenchyma. The patient had ACTH serum level: 499,0 pg/mL (normal < 46,0 pg/mL); cortisol serum level: <1 µg/dL (normal 5 to 25 µg/dL).

References: Department of Radiology, Clinics Hospital, University of São Paulo, SP, Brazil
Fig. 2: Testicular adrenal rests tumors. 16-year-old male with poorly controlled CAH was referred for evaluation due to testicular pain and bilateral palpable mass. (A) Axial and (B-C) sagittal US images demonstrated bilateral nodular hypoechogenic intratesticular masses, located around the mediastinum.

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In some cases, few of the lesions can present acoustic shadowing. (Fig 3)
Fig. 3: Testicular adrenal rests tumors. 17-year-old male with CAH was referred for routine evaluation. At physical examination testicular size was normal, there was no palpable masses. (A) Axial and (B-C) sagittal ultrasonographic images demonstrated bilateral nodular hypoechogenic intratesticular masses, located around the mediastinum testis that determine slight acoustic shadowing.  

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Testicular adrenal rests tumors usually present a slightly increased vascularity compared to the normal testicular parenchyma at color-Doppler sonography. (Fig 1, 4 and 5)
**Fig. 4**: Testicular adrenal rests tumors. 16 years old male with CAH. (A) Axial and (C) sagittal ultrasonographic images demonstrated bilateral nodular hypoechogenic intratesticular masses, characteristically located around the mediastinum testis. At (B) color-Doppler evaluation, the TART have slightly increased vascularity compared to the adjacent normal testicular parenchyma.

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**Fig. 5:** Testicular adrenal rests tumors. 3 years old male with CAH. (A) Axial and (B) sagittal ultrasonographic images demonstrated bilateral nodular hypoechogenic intratesticular masses, characteristically located around the mediastinum testis. At (C) power-Doppler US evaluation, the testicular adrenal rests tumors show increased vascularity compared to the adjacent parenchyma.

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Although the testicular adrenal rests tumors are usually hypoechogenic, lesions larger than 2 cm can be heterogeneous, sometimes hyperechogenic, and may present calcifications. (Fig 6)
Fig. 6: Testicular adrenal rests tumors. 24 years old male with CAH. (A) Axial and (C-D) sagittal US images obtained in three different locations demonstrated bilateral nodular heterogeneous intratesticular masses, with calcifications.

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Testicular adrenal rests tumors have no malignant features. However, because of the localization near the mediastinum testis, compression of the seminiferous tubules might lead to significant damage of the surrounding testicular tissue, leading to infertility. (Fig 7)
**Fig. 7:** Ectasia of the rede testis in a patient with CAH and testicular adrenal rest tumors

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Testicular adrenal rest tumours in CAH might impair both spermatogenesis and endocrine testicular function, resulting in infertility. Poor controlled disease, resulting in elevated ACTH serum levels is thought to induce the TART growth. However, the association of TART and disease control is not entirely understood. Some individuals might present increase in TART despite good disease control parameters. (Fig 9 and 10)
**Fig. 9**: Testicular adrenal rests tumors. 16 months old male with CAH. (A) Axial and (B) sagittal images demonstrated bilateral nodular heterogeneous intratesticular masses, near the mediastinum testis, with slightly increased vascularity on color-Doppler evaluation. ACTH serum level: 40,0 pg/mL (normal < 46,0 pg/mL); cortisol serum level: <1 µg/dL (normal 5 to 25 µg/dL).

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Fig. 10: US evaluation of the same individual depicted in Fig 9, after two years, shows a significant increase in size of the bilateral lesions, despite the good clinical treatment control. ACTH serum level: 31.0 pg/mL (normal < 46.0 pg/mL); cortisol serum level: 1.2 µg/dL (normal 5 to 25 µg/dL).

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MR Imaging

Testicular adrenal rests tumor appear homogeneous on both T1- and T2-weighted MR images. TART are commonly isointense, eventually hyperintense, to normal testicular tissue on T1-weighted MR images and hypointense on T2-weighted MR images. TART are isointense to muscle tissue on both T1- and T2-weighted MR images, similar to MR appearance of the normal adrenal glands. In most cases, these lesions enhance after gadolinium administration.

Currently the diagnosis of CAH is generally established by neonatal screening programs. However, in individuals with milder forms of the disease the accurate diagnosis may not be established until late childhood or even early adulthood. (Fig 11 and 12)
**Fig. 11:** Testicular adrenal rests tumors. 45-year-old male referred for evaluation due to testicular palpable right testicular mass. US evaluation demonstrated bilateral heterogeneous, intratesticular, ill-defined masses, located around the mediastinum testis, that determine slight acoustic shadowing.

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Fig. 12: Testicular adrenal rests tumors. MR imaging of the same individual depicted in Fig 11 (A) On axial T1-weighted MR images the lesions are isointense relative to the testicular parenchyma. (B and D) Axial and sagittal T2-weighted MR image show a lobulated intratesticular lesion that is hypointense compared with normal testicular parenchyma. (C) Enhanced sagittal T1-weighted MR image shows slight homogeneous enhancement of the intratesticular lesions.

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The main differential diagnosis is with Leydig cell tumor and lymphoma.

Images for this section:

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**Fig. 7:** Ectasia of the rede testis in a patient with CAH and testicular adrenal rest tumors.
Fig. 8: Strain elastography in testicular adrenal rests tumors. Elastogram shows predominately green color inside the TART, similar to the normal testicular parenchyma (red is soft and blue is stiff).
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Conclusion

- Imaging plays an important role in the detection and surveillance of testicular adrenal rest tumors.
- US and MR imaging features are characteristic in the context of elevated ACTH serum level.
- Familiarity with the imaging appearance of the testicular adrenal rests tumors will help to establish an accurate diagnosis and avoid unnecessary invasive procedures.

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References


