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Diagnosis of Testicular Adrenal Rest Tumors on Ultrasound : A Study of 01 Case Report

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Abstract: *Testicular adrenal-like tissue are found in ACTH hypersecretion syndromes, most often in congenital adrenal hyperplasia CAH attributable to a 21-hydroxylase deficiency. They are detected clinically or systematically on ultrasound scan, generally for young adults. In sonography, they appear as confluent hypoechoic masses, often bilateral, which developed into region of the testicular hilum. They get high in the late stage suggestive damping aspect. In Doppler color they have regular vascular architecture. They generally regress or stabilize after replacement treatment with glucocorticosteroids. Biopsy or seminal vein samples they are not reliable. MRI does little more than ultrasound. Diagnosis involves observation or, ultimately, an orchiectomy. Here we presented a case of a patient with bilateral testicular masses and an adrenal mass which have a common cause in the setting of congenital adrenal hyperplasia.*

Keywords: Testicular-adrenal resting tumors. congenital adrenal hyperplasia. Imaging.

INTRODUCTION

Mainly adrenal testicular resting tumors (TART) occur in patients with congenital adrenal hyperplasia (CAH), one of the most common congenital adrenal gland disease, attributable to a 21-hydroxylase deficiency.
Due to their resemblance to the adrenal glands' tissues, these lesions were originally called "testicular adrenal rest tumors".
Classic therapy CAH forms consist of a glucocorticoid throughout life substitution. Decreased fertility is a serious, long-term complication.
Here we presented a case of a patient with multiple testicular masses and an adrenal mass which have a common cause in the setting of congenital adrenal hyperplasia.

PATIENT AND OBSERVATION

This is a 10-year-old child of a second-degree consanguineous marriage of a 31-year-old mother, and a 44-year-old father, farmer. Admitted to the emergency room for sudden testicular pain, with suspected testicular torsion.
The patient had the following history: Congenital adrenal hyperplasia on probable 21 Hydroxylase deficiency, revealed by a sexual differentiation anomaly in the neonatal period, put on Hydrocortisone 20 mg/d replacement therapy, lost to follow-up 3 years ago with poor therapeutic compliance. He had precocious puberty since the age of 3 years and was operated for vulviform hypospadias. The interrogation notes the installation of atrocious testicular pains, evolving in a context of fever and alteration of the general state, with suspected testicular torsion.
The clinical examination found a patient weighing 35 kg (+1.5 DS) for a height of 148 cm (+2.5 DS), pubescent Tanner stage 4, melanoderma, without dysmorphic syndrome.
Examination of the external genitalia: Male type, presence of pubic hair, enlarged testicle, painful with nodular aspect on palpation; no other anomalies.
The patient underwent a testicular ultrasonography. It revealed a markedly heterogeneous appearance of both testes. The testicules were enlarged bilaterally with lobulated contours (*Figure 1-2*), nodular hypoechoic mass-like areas, confluent, demonstrating posterior acoustic shadowing. This aspect was centered around the testicular hilum, with preservation of the

mediastinum. Doppler showed mostly peripheral vessels, with hypervascular pseudo-nodules. There was no sign of testicular torsion bilaterally. (*Figure 3*)

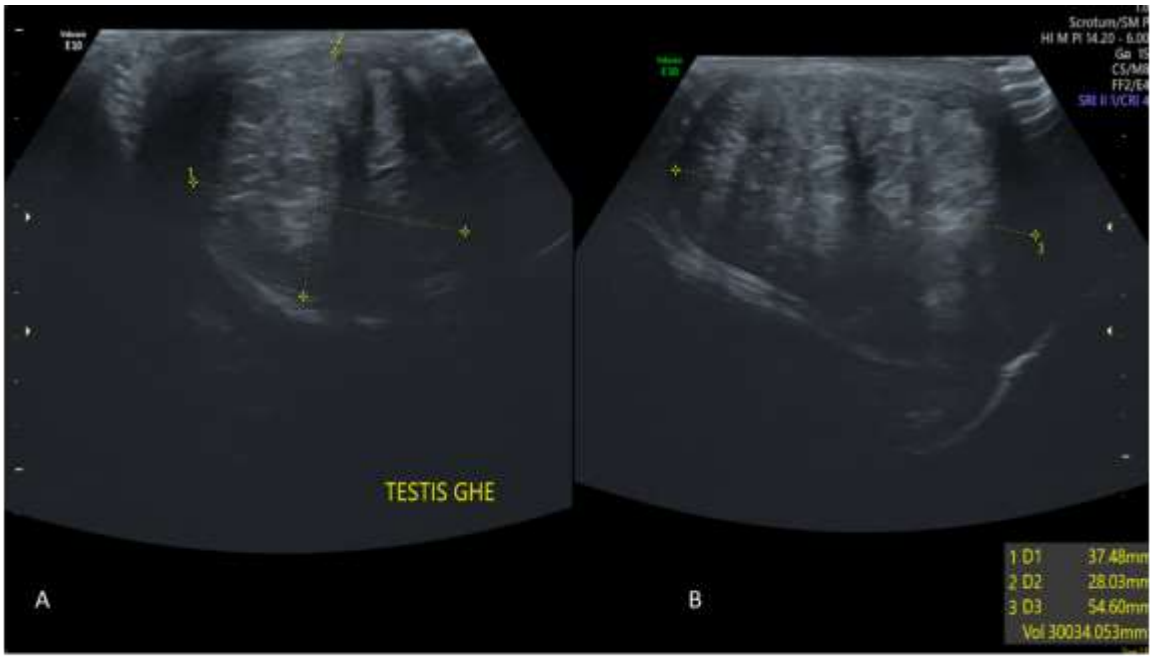

Figure 1: *Ultrasound images of the left testis in the transverse (A) and sagittal (B) planes show an enlarged testicule, with diffuse, heterogeneous appearance.*

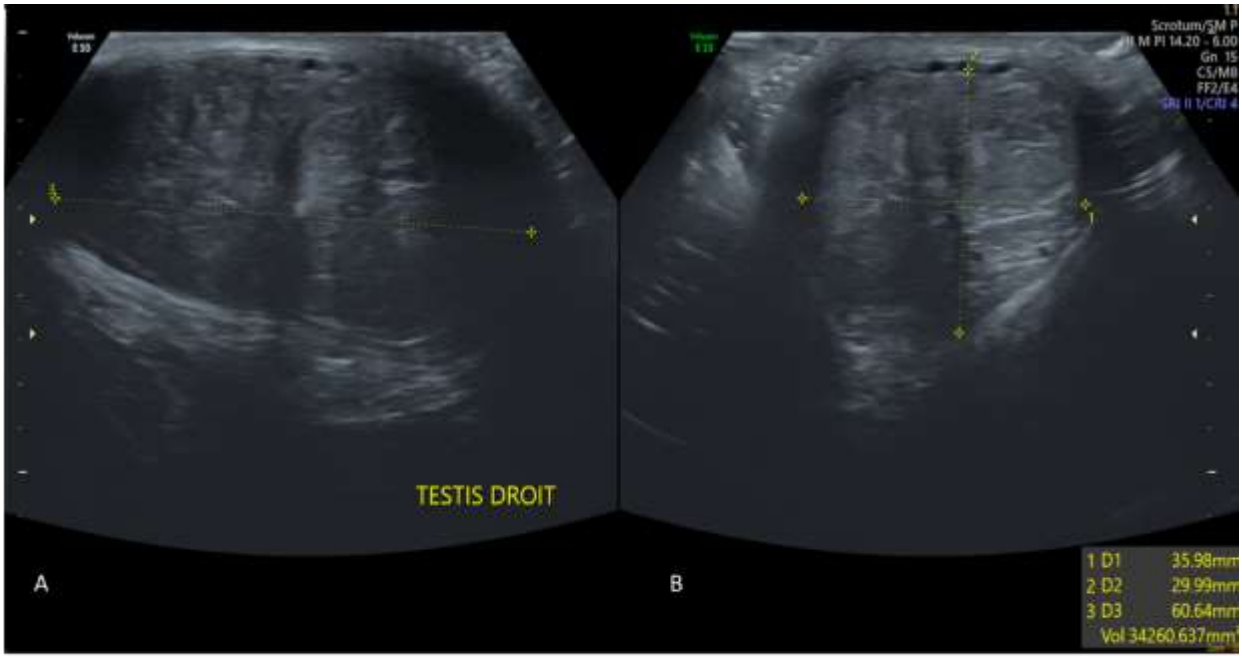

Figure 2: *Ultrasound images of the right testis in the transverse (A) and sagittal (B) planes show an enlarged testicule, with diffuse, heterogeneous appearance.*

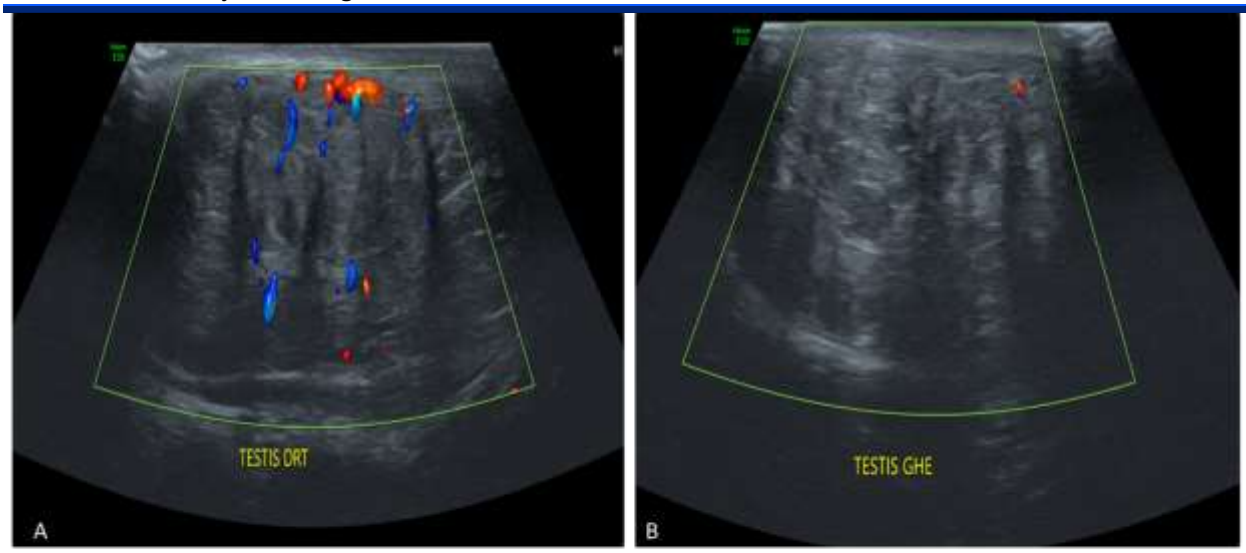

Figure 3 (A-B): Markedly heterogeneous appearance of both testes with nodular hypoechoic mass-like areas, confluent, demonstrating posterior acoustic shadowing. This aspect was centered around the testicular hilum, with preservation of the mediastinum.
Doppler showed mostly peripheral vessels, with hypervascular pseudo-nodules.

Given the clinical history of congenital adrenal hyperplasia, an abdominal ultrasound complement was performed, objectified a right adrenal mass suggesting first an adrenocortical cancer; (*Figure.4*)

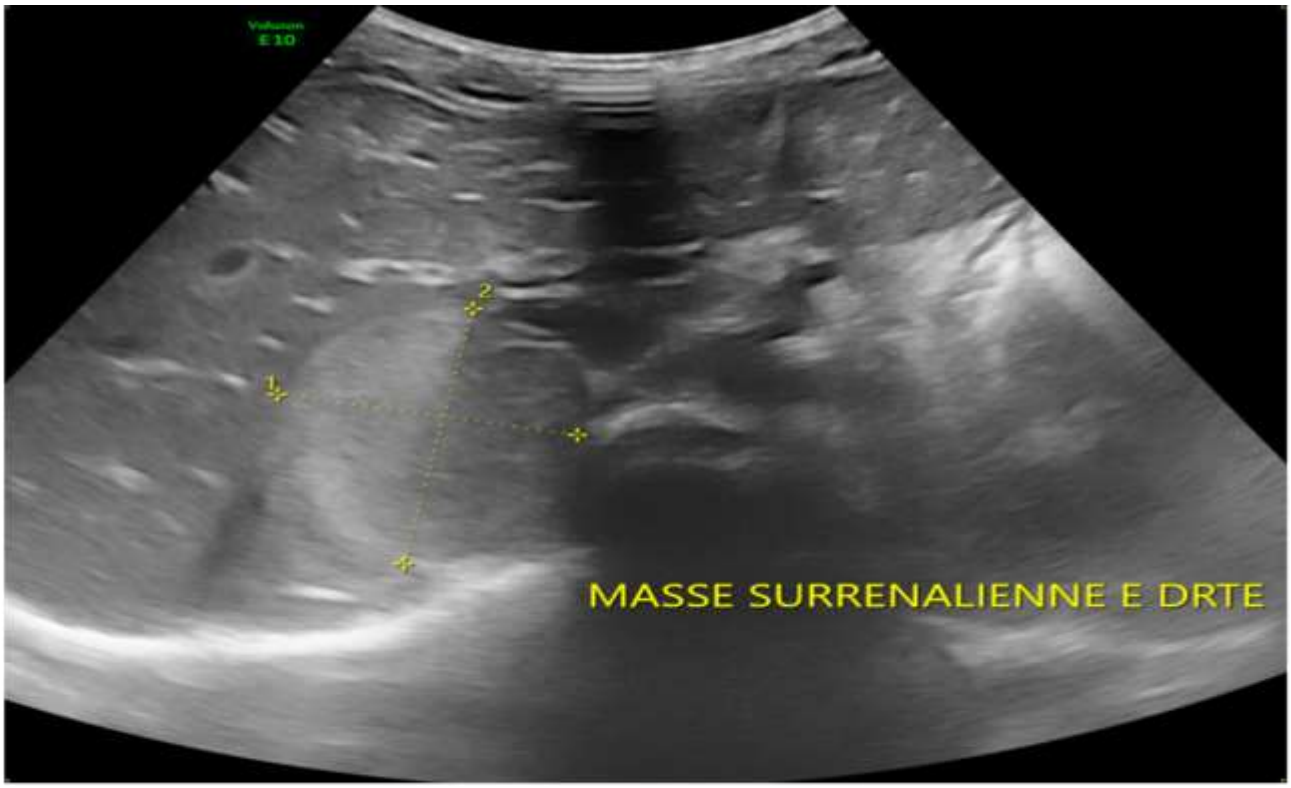

Figure 4: Right adrenal mass, with smooth contours, iso echoic, homogenous, measuring 03 cm.

Laboratory tests showed: sterile ECBU, CRP: 319, serum cortisol: 376 ng/ml, testosterone:0.60 ng/ml LH 0.01 mUI/ml ; 17 alpha hydroxyprogeterone : 4.80 ; FSH : 0.33 ; Delta 4 androstenedione : 0.50 ng/ml , **Karyotype** : 46, XY .

DISCUSSION:

Testicular adrenal rest tumors (TART) are benign tumors that occur secondary to longstanding stimulation of ectopic adrenal tissue within the testis of patients with CAH. These tumours can be already found in childhood and puberty, with a frequency of 18.3% to 29% in studies with children ([i],[ii]).

Clinically, TARTs are usually asymptomatic, sometimes they are responsible for embarrassing scrotal heaviness or pain. They are most often bilateral, 83% according to Rutgers (19), and develop gradually, especially in patients with endocrinologic disorders. When felt, they are usually firm and may be somewhat imposing for a malignant tumor. ([iii],[iv]).

Due to the often-asymptomatic nature of TART, the diagnosis is usually not performed until the tumors are left behind large enough to be palpated or induced mechanical obstruction. ([v],[vi],[vii])

Gonadal ultrasound is necessary even in asymptomatic patients with negative clinical exploration ([viii],[ix]). In patients diagnosed with CAH, TART must be diagnosed earlier during routine screening tests, and ultrasound is the best, most cost-effective imaging TART diagnosis modality. In fact, TART is still too often only found in the infertility stage.

The ultrasound imaging features of TART show a wide variety of features, but tend to be multiple bilateral masses located along the mediastinum testis of varying echotextures with vascular architecture preserved on doppler imaging. The masses are usually well marginated. They are usually not associated with calcifications. [x]

However, on MRI, TART is isointense to the testicular parenchyma on T1 weighting and isointense or slightly hypointense on T2 weighting. They are enhanced after injection of gadolinium. ([xi],[xii])

MRI has not been shown to be superior by compared to ultrasound for differential diagnosis between TART and germinal tumors. Its only advantage would be better visualization of tumor boundaries ([xiii]), which can sometimes be useful with orchiectomy be useful with partial orchiectomy is considered ([xiv]).

The main diagnostic considerations for TART are malignant tumor (both primary and metastatic diseases), infections and sarcoidosis.

Histologically, TARTs are similar in appearance and morphology to adrenocortical tissue, but may sometimes be mistaken for malignant Leydig cell tumors. Immunohistochemical stains remain helpful on tissue evaluation to help discriminate between the two entities.

Based on the histological appearance of TART and the surrounding testicular parenchyma and the clinical observations, the development of TART was divided in five different stages by ***Claahsen-van der Grinten HL et al.([xv])*** (Figure.5)

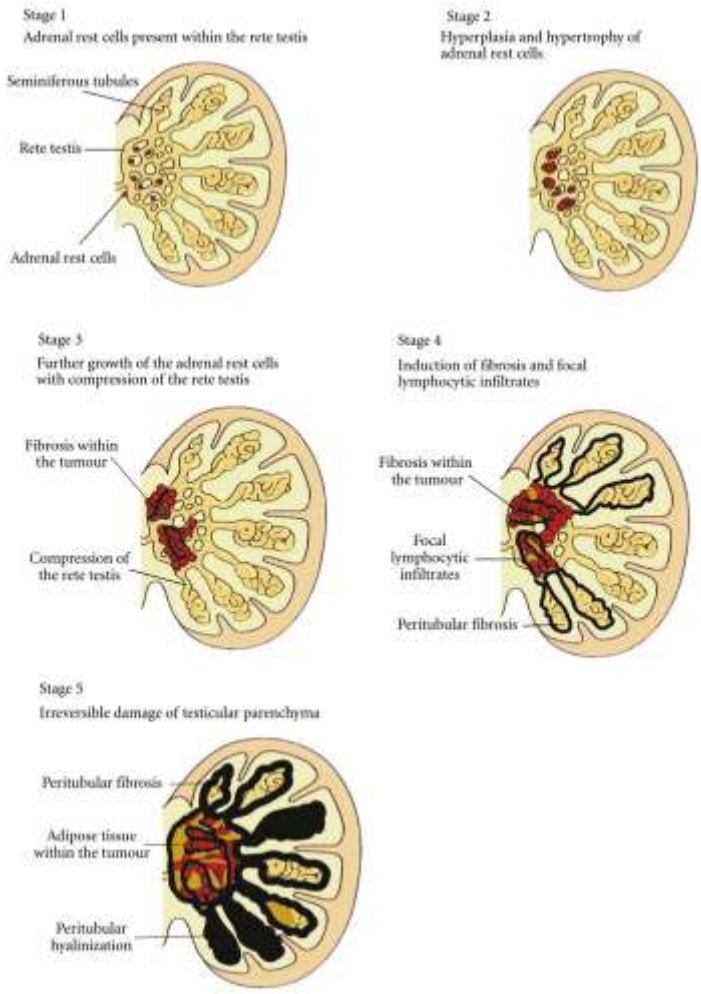

Figure 5: Proposed staging of TART by the Claahsen-van der Grinten HL et al.

Claahsen-van der Grinten, H.L., Hermus, A.R.M.M. & Otten, B.J. Testicular Adrenal Rest Tumours in Congenital Adrenal Hyperplasia. *Int J Pediatr Endocrinol* **2009**, 624823 (2009). https://doi.org/10.1155/2009/624823

As they are benign lesions, TARTs are typically treated with medications, with the administration of adequate amounts of exogenous glucocorticoids. Rarely, patients may be considered for further treatment including partial or total orchiectomy [xvi,xvii].
The person concerned should be encouraged to perform a spermogram and freeze the gametes for subsequent in vitro fertilization if necessary (xviii).

CONCLUSION:

In conclusion, it is important for both clinicians and radiologists to become familiar with the congenital adrenal hyperplasia CAH and testicular adrenal rest tumors TART.
Boys with congenital adrenal gland hyperplasia should benefit from regular routine scrotal ultrasound examinations from childhood. If testicular nodules are found, the diagnosis of TART is by far the more likely, especially if it is a prepubertal boy if the crisis is bilateral, and if the appearance the echo-Doppler is evocative. Regression of the abnormalities will provide the diagnosis.
MRI have an interest to guide possible procedure of partial orchiectomy.

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