An unusual Testicular Adrenal Rest Tumor localization in a 15-year-old boy with congenital adrenal hyperplasia.

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Introduction

Testicular adrenal rest tumor (TART) is the most important cause of infertility in male with congenital adrenal hyperplasia (CAH). TART is a benign tumor, mainly bilateral (75-80%), usually diagnosed in under-treated CAH male with hypercorticotropinemia, which generally regresses after glucocorticoids therapy adjustment. However, it may determine an irreversible damage by compression and toxic-paracrine effects on the surrounding testicular tissue.
At ultrasound (US) examination, TART appears as hypoechoic, clearly delineated, rich and regular in vascularization, testicular lesions usually localized close or inside the testicular hilum. We report the first documented case of unusual epididymal localization of TART in an adolescent with salt-wasting (SW) CAH.

Case report

A 15-year-old Caucasian boy was diagnosed with SW CAH (Intron-2-splice mutation and 8-bp-deletion) because of an adrenal crisis at the age of 1 month. He was regularly followed up through biochemical tests and testicular US evaluations and treated lifelong with hydrocortisone and fludrocortisone acetate until the age of 14 when a progressive deterioration of disease control due to lack of compliance to therapy was reported (Tab.1). At the age of 15, scrotal US examination, demonstrated, for the first time, multiple, homogeneous and well-circumscribed hypoechoic lesions impairing both testes ranging between 3 mm to 9 mm, most of them located along mediastinum testis, and two other nodular lesions in both epididymis heads, the greater of which measuring 7 mm in size in the right one. Testicular and epididymal lesions showed increased intra and perilesional vascularization, associated with regular and linear caliber of the vessels coursing through the lesions at power Doppler evaluation, higher stiffness values compared to testicular parenchyma at strain elastography (SE) assessment, and low intensity signal in the T2 Weighted MRI images (Fig.1). Eight months after TART's diagnosis and about 6 months after patient restarted glucocorticoid therapy, a reduction in size of testicular and epididymal lesions was documented at US.

Tab.1- Follow-up of laboratory tests and testicular volume at ultrasound

Age (years)		12.7	13.7	13.9	14.3	15 *	15.3	15.8	N
ACTH (pg/ml)		113	>1250	488	827	1057	440.7	430	6 - 40
17-OHP (ng/ml)		14.9	43.6	44	89	87	33	27	0.3 - 2.2
Delta4-A (ng/ml)		0.8	15.1	4.5	8	17	8	7.6	0 - 2.9
Renin (uUI/ml)			35.9	43.1	16.5	16.9	13.5	14.4	3.6 - 20
Testicular volume (ml)	Right					24.5		24.3	
	Left					22.8		22.3	

TART diagnosis (*), normal hormonal range (N), 17-hydroxyprogesterone (17-OHP), Delta4-androstenedione (Delta4-A)

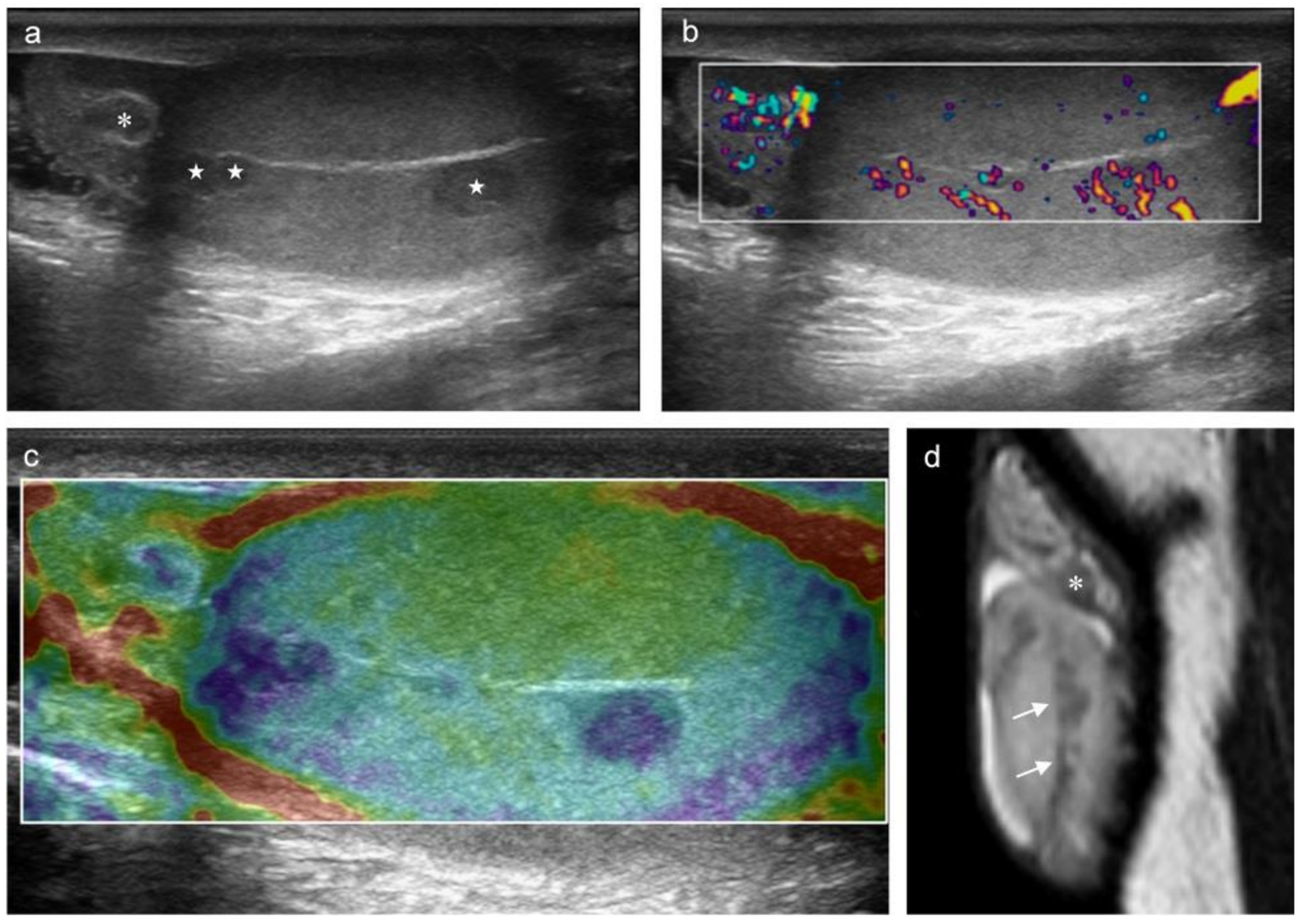

Figure 1 – Ultrasound and Magnetic resonance imaging of TART

a) b-mode ultrasonography image of the right testis;
b) Power-Doppler ultrasonography image of the right testis;
c) Strain Elastography ultrasonography image of the right testis;
d) T2W MRI image of the right testis.
Images showed multiple intratesticular solid masses, located along the mediastinum testis (stars in a and arrows in d) and a single mass into the head of epididymis (astersisk in a and d).

Conclusions

This is the first documented case of epididymal localization of TART in an adolescent with SW CAH. The diagnosis of TART should be always considered in CAH male with testicular lesions and an epididymal localization should be encountered. In those patients, testicular US screening should be performed regularly, at least every two years in early childhood and annually in the peripubertal period, or even more frequently in patients with lack of compliance to glucocorticoids therapy, even in absence of suggestive symptoms.

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