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Large testicular adrenal rest tumours (TART) in a patient with congenital adrenal hyperplasia: a consequence of poor drug compliance

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Introduction

Adrenal Rest Cells first described in 1940 are a group of cells trapped within the developing gonad during foetal development.[1] Usually less than 5 mm in size, found in the testis and surrounding tissue in 7.5-15% of newborn and in about 1.5% of adults.[2] Testicular adrenal rest tumours (TART) are benign corticotrophin-dependent tumours that occur in males with congenital adrenal hyperplasia (CAH). We present a patient with bilateral large TART as a consequence of poor compliance to treatment and follow-up.

Case

A 25 year old gentleman presented to the endocrine clinic in 2009 with a history of tiredness, reduced libido and bilateral large testicles, which he wanted surgically removed. He had been diagnosed with CAH during his antenatal period and commenced on steroid replacement therapy soon after birth. However, he stopped taking all medications in 2000 because of family issues and although his testicles were felt to be lumpy in 2002, further follow-up/investigation was difficult. He was well with normal secondary sexual features, but his testicles felt hard and three times the normal size.

Investigations

His biochemical test results are shown in the Table 1. An ultrasound confirmed large testicles. He was commenced on steroid replacement therapy* and referred for orchidectomy and replacement prosthesis. Both testicles measured 8.5 x 5 x 4cm in size with no recognisable testicular parenchyma (stage 5, Figure 1): the histology was characteristic of TART. His serum testosterone level fell to the lower limit of the normal range post-surgery. He has not attended clinic since then, despite multiple attempts to contact him.

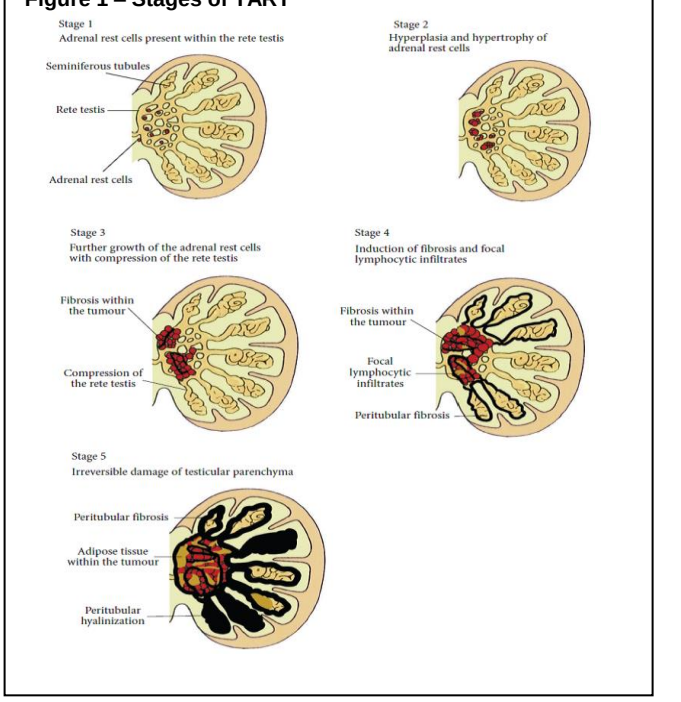
Table 1

Biochemical test	Results	Normal reference range
17-OH-Progesterone	>152	<13 nmol/L
Testosterone	50.4	10-38 nmol/L
Corticotropin (ACTH)	139	0-50 ng/L
Dehydroepiandrosterone	12	1.6-11 µmol/L
Follicle stimulating hormone	<1	1-14 U/L
Luteinising hormone	<1	1-9 U/L
Prostate specific antigen	0.27	<2.5 µg/L
0-minute cortisol (pre-Synacthen)	327	----------
30-minute cortisol (post-Synacthen)	354	>550 nmol/L
Sodium	144	133-146 mmol/L
Potassium	4.2	3.5-5.3 mmol/L

*Was commenced on:

Hydrocortisone 5mg bd
Dexamethasone 0.25mg nocte

Figure 1 – Stages of TART[2]



Conclusion

Although not malignant, TART can result in irreversible damage of testicular tissue and infertility. Treatment with glucocorticoid replacement therapy can stabilise or repress growth of these tumours. This case highlights the importance of compliance to treatment and follow-up to prevent testicular damage as a result of TART in patients with CAH.

References

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