TESTICULAR ADRENAL REST TUMOUR IN CHILDREN WITH CLASSICAL CONGENITAL ADRENAL HYPERPLASIA: A CASE SERIES

H.P. CHENG[1], J.S.L. WONG[1], M.S. HASSIM[2], N.A. MAT AMIN[1], S.T. TEOH[1], C.G. GAN[1], L.A. ANAND[1], N.M. SELVEINDRAN[1], J.Y.H. HONG[1]
[1]Paediatric Endocrine unit, Putrajaya Hospital, Malaysia
[2]Radiology unit, Putrajaya Hospital, Malaysia

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INTRODUCTION

Testicular adrenal rest tumour (TART) is a complication in males with congenital adrenal hyperplasia (CAH). TART could lead to infertility by causing obstructive azoospermia[1]. However, the prevalence, risk factors and treatment in children are not clear.

AIM

1) To identify the incidence of TART and risk factors in patients with classical CAH in our centre.

2) To study the clinical characteristics, hormonal profile, treatment interventions and outcome of patients with TART.

METHOD

Testicular ultrasound were performed in 16 patients with poorly controlled CAH (mean age 9.87 ± 3.11, range 4.67-16yrs).

Clinical characteristics, hormonal profiles and treatment were compared between patients with and without TART.

RESULTS

TART lesions were detected by ultrasound in 6 patients (37.5%). The lesions were bilateral in all patients, mean diameter of 0.3cm, hypoechoic in nature and scarce in vascularity. Interestingly, testicular microlithiasis was found coincidently in 1 patient. Increased frequency of testicular microlithiasis in CAH is postulated due to multifactorial cellular degeneration, which includes TART[2].

There was no significant difference in height SDS and BMI SDS in patients with and without TART. TARTs were detected in 60% of patients who had attained final height. Only 1 patient had palpable nodules. Clinical evaluation had as sensitivity of 16.7% when compared to ultrasonographic evaluation in our study, as majority of the lesions were small.

5 patients developed peripheral precocious puberty and 2 patients evolved into central precocious puberty. Although poor hormonal control has a pivotal role in the pathogenesis of TART, one patient developed TART despite good control. This has also been demonstrated in other studies indicating the likelihood that other factors are also involved in the pathogenesis of TART[3].

All had intensified hydrocortisone regimen after detection of TART (mean dose 17mg/m^2/day) for 2-4 weeks, however only two (33%) had resolution on repeated ultrasound. Our data showed a lower rate of disappearance or resolution of TART on follow up ultrasound compared to adult studies despite a 25-30% increment of hydrocortisone dosage[4]. This could be contributed by the poor compliance reported most of our patients.

Fig 1: Ultrasound showed homogenous hypoechoic lesions with clear boundary on both testes.

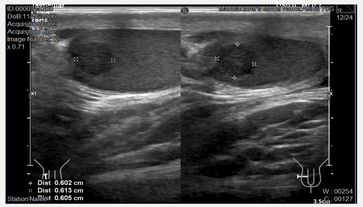

Table 1. Clinical characteristics of patients with TART

Patient	Age of CAH diagnosis, years	Age of TART diagnosis, years	17OHP (nmol/L)	Hydrocortisone dose (mg/m2/day)	Resolution of TART
1	0.09	4.67	>60.6	18.0	Yes
2	0.3	6.00	8.8	14.5	Yes
3	0.09	13.08	141.6	18.0	No
4	0.09	13.83	572.0	18.4	No
5	3.50	8.33	246.2	17.9	No
6	0.09	11.75	614.9	16.9	No

Fig 2: Coindicental bilateral microlithiasis in one patient with TART.

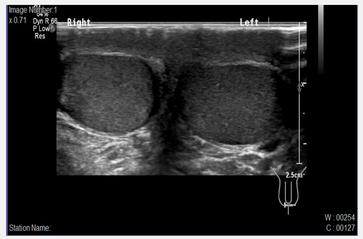

Table 2. Patients with and without TART: Comparison of clinical characteristics

	With TART (n=6)	Without TART (n=10)	p value
Age, years	9.61 ± 3.83	10.03 ± 2.81	0.806
Weight SDS	1.29 ± 0.89	0.29 ± 1.43	0.150
Height SDS	0.76 ± 1.06	-0.24 ± 1.69	0.213
BMI SDS	1.18 ± 0.86	0.82 ± 0.98	0.471
Bone age advancement	4.89 ± 2.95	1.28 ± 3.10	0.037
17OHP	322.22 ± 238.87	149.61 ± 201.20	0.143
Hydrocortisone	15.10 ± 2.42	14.14 ± 1.75	0.371

CONCLUSIONS

Ultrasound surveillance for TART in males should be performed in childhood, especially in those with poor disease control and at peri-pubertal age.

Larger studies should be conducted to explore the risk factors and therapeutic modalities in TART.

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CONTACT INFORMATION

Paediatric Endocrine Unit,
Putrajaya Hospital,
Jalan P9, Presint 7, 62250 Putrajaya, Wilayah Persekutuan Putrajaya, Malaysia.
Email: hpj.paedsendo@gmail.com

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