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Testicular masses in congenital adrenal hyperplasia: Using ^{123}I-MIBG scintigraphy to support the diagnosis of testicular adrenal rest tumours

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Abstract

This case demonstrates the use of ^{123}I-MIBG scintigraphy in the diagnosis of testicular adrenal rest tumours (TART) in an adult with classical congenital adrenal hyperplasia (CAH). TART are common in CAH patients, with MIBG scanning offering a new imaging modality to potentially help verify the diagnosis and prevent invasive investigation.

Level of evidence: Level 5 case report

Keywords

Testicular adrenal rest cell tumours, congenital adrenal hyperplasia, testicular mass, MIBG scintigraphy

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Introduction

Congenital adrenal hyperplasia (CAH) is an autosomal recessive disorder that is commonly caused by 21-hydroxylase deficiency in the adrenal gland, resulting in impaired corticosteroid synthesis and excess androgen production.[1,2] Via negative feedback mechanisms, pituitary adrenocorticotropic hormone (ACTH) production is increased stimulating adrenal gland hyperplasia.[1]

During embryonic development, the adrenal glands and the gonads both derive from the urogenital ridge.[2] Aberrant adrenal tissue may travel with the testes during their descent into the scrotum, occurring in 7.5%–15% of normal neonates.[3] High ACTH levels in CAH cause the adrenal tissue remnants to become hyperplastic, developing into testicular masses. Testicular adrenal rest tumours (TART) are reported to be more common in patients with suboptimal hormonal replacement.[3,4]

TART are present in up to 94% of patients with 21-hydroxylase deficiency.[1,2] TART are benign and located in the *rete testis*. Clinically, these lesions are usually bilateral (80% cases), synchronous and initially become palpable at 2 cm in size.[1,2,5] Ultrasonography is the radiological investigation of choice to detect and monitor these tumours; however, it is non-specific and there are no recognised screening guidelines.[4]

Case report

A 37-year-old male was referred to our urology outpatient department regarding bilateral testicular masses confirmed on ultrasound as part of surveillance screening for classical CAH. On examination male genitalia appeared normal; however, palpable bilateral testicular lumps were detected on palpation measuring 1 cm to 2 cm in the superoposterior aspect.

Biochemical testing revealed normal serum tumour makers, with an alpha fetoprotein of 3 ug/l (normal < 12 ug/l)

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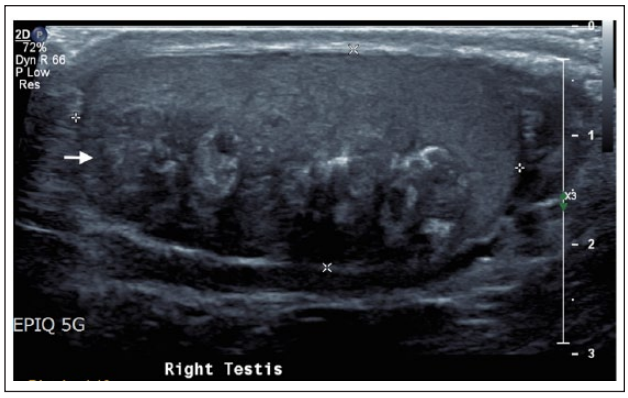

Figures 1. Ultrasound image showing a testicular lesion of altered heterogeneous echotexture within the right testis (indicated by white arrow) measuring 20 × 35 × 15 mm.

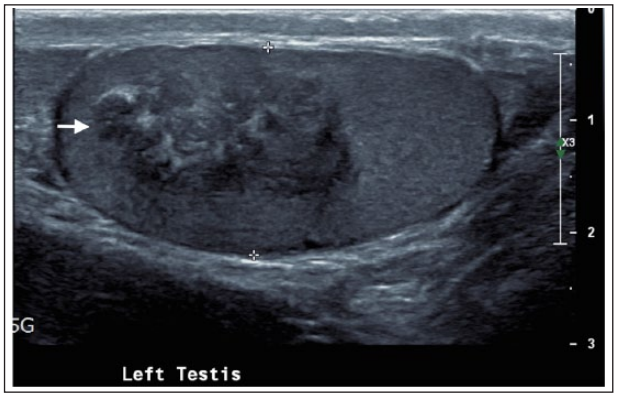

Figure 2. Ultrasound image showing a testicular lesion of altered heteogenous echotexture within the left testis (indicated by white arrow) measuring 17 × 16 × 30 mm.

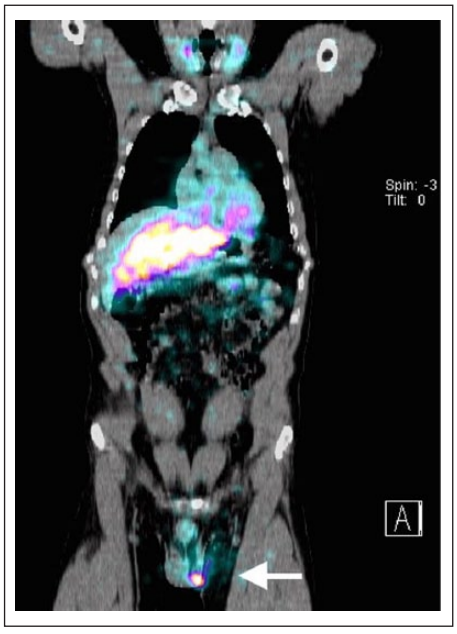

Figure 3. Coronal whole body SPECT CT image scanned at four hours post MIBG injection. This images shows focal MIBG avidity in the scrotum (white arrow), which indicates ectopic adrenal medullary tissue in the scrotum.

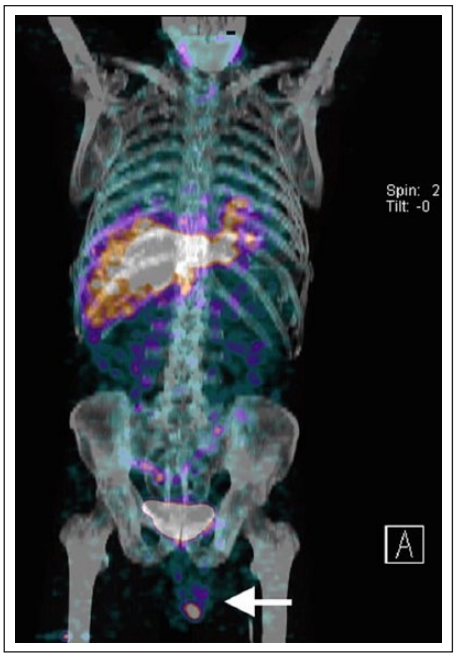

Figure 4. Coronal whole body SPECT CT image scanned at four hours post MIBG injection. The white arrow indicates focal ectopic MIBG avidity in the scrotum.

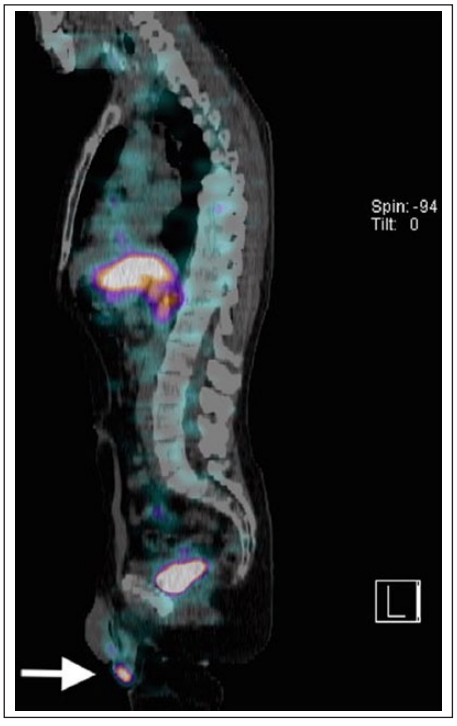

Figure 5. Sagittal whole body SPECT CT image scanned at four hours post MIBG injection. The white arrow indicates focal extopic MIBG avidity in the scrotum.

and human chorionic gonadotrophin levels of < 2 IU/l (normal <2 IU/l). ACTH level was elevated at 730 ng/l (normal 5 ng–50 ng/l).

On initial ultrasound (see Figures 1 and 2) there were lesions of heterogeneous altered echotexture within the

body of each testis, on the right measuring 20 × 35 × 15 mm and on the left measuring 17 × 16 × 30 mm. These were associated with increased vascularity. On interval ultrasound imaging six weeks later the bilateral testicular lesions were unchanged in size and appearance.

Whole-body meta-iodobenzylguanidine (MIBG) scans using 233 Mbq ^{123}I-MIBG was performed with imaging at four and 24 hours with single-photon emission computed tomography (SPECT) of the chest, abdomen and pelvis. Imaging performed at four hours demonstrated focal increased MIBG activity in the scrotum (see Figures 3–5). The MIBG scan thus supported the presence of adrenal medullary tissue within the scrotum, corresponding with the lesions detected on ultrasound.

Discussion

To our knowledge ^{123}I-MIBG scintigraphy has not been previously reported in diagnosing TART. A literature search of PubMed and Medline databases using the search term "testicular adrenal rest tumour AND MIBG" returned no results. Currently, ^{123}I-MIBG scanning is most commonly used in the diagnosis of phaeochromocytoma. MIBG uptake occurs in the plasma membrane norepinephrine transport system and is concentrated within the sympathomedullary system.[6]

Accurately diagnosing TART early is important. Primarily, it is essential to exclude sinister causes of bilateral testicular masses, namely malignant Leydig cell tumours.[1,7] Despite the likelihood of benign disease in these patients, surgical removal may be performed with significant consequences. Ali et al. reported a case of bilateral orchidectomy in a 15-year-old patient due to diagnostic uncertainty and concern for malignancy.[5]

MIBG scanning in these scenarios could provide a useful adjunct to standard investigations, as it is specific for sympathomedullary tissue and thus may confirm the presence of ectopic adrenal tissue in the scrotum. In conjunction with normal tumour markers, this investigation may reassure the clinician of benign disease and help to reduce patient anxiety regarding malignancy. Additionally, MIBG scans may replace further investigations and reduce the associated financial and psychological impacts of ongoing tests for patients.

Conclusion

This case used ^{123}I-MIBG scintigraphy to support the diagnosis of TART in this patient and highlights the potential utility of this investigation in aiding in diagnosis of this condition. A positive MIBG scan in our case substantiated the diagnosis of adrenal rest cell tumours, negating further intervention at this stage and supported surveillance of the testicular lesions rather than operation.

Conflicting interests

The Authors declare that there is no conflict of interest.

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Ethical approval

Informed consent for patient information and images to be published was provided by the patient. Research protocol was approved by the Darling Downs Hospital Health Service, Toowoomba Hospital, Toowoomba QLD 4350, Australia.

Informed consent

Informed consent was obtained from the patient for his anonymized information (including images) to be published in this article.

Guarantor

LEH.

Contributorship

ARS and LEH conceived the study. All authors researched the relevant literature. The initial and subsequent drafts of the manuscript were written by LEH. All authors reviewed and edited the manuscript and approved the final version of the manuscript.

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