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Testicular Adrenal Rest Tumour (TART) or Testicular Malignancy: A Clinical Dilemma

Moni Roy, Ashish Kumar Roy, Tulika Chatterjee, Saurabh Bansal
Department of Internal Medicine, University of Illinois College of Medicine at Peoria and OSF Saint Francis Medical Center, Peoria, Illinois, USA

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ABSTRACT

Testicular adrenal rest tumour (TART) is a known entity in patients with congenital adrenal hyperplasia. An adult patient presenting with testicular enlargement raises a concern for malignancy and this creates a diagnostic dilemma between non-malignant conditions such as TART versus testicular malignancy. We describe a case where the patient underwent orchiectomy due to clinical concern for malignancy but, retrospectively, this outcome could have been prevented by medical treatment. This case emphasises the need to learn from errors. There is a need to increase awareness of the condition among medical professionals to reduce the chances of unnecessary surgical intervention.

LEARNING POINTS

- To recognize testicular adrenal rest tumour (TART) as cause of testicular enlargement in adult patients with congenital adrenal hyperplasia.
- To differentiate TART from other types of testicular malignancy.
- Consider medical treatment with exogenous glucocorticoid and mineralocorticoid replacement to prevent unnecessary surgical intervention.

KEYWORDS

Testicular adrenal rest tumour, congenital adrenal insufficiency

CASE DESCRIPTION

A 45-year-old man initially presented to our hospital with pleuritic chest pain. Computed tomography (CT) of the chest ruled out a pulmonary embolism but showed incidental findings of bilateral multiple adrenal nodules as shown in Fig. 1. The patient reported a history of congenital adrenal hyperplasia and inconsistent use of steroid replacement since his teenage years. Interestingly, he reported no prior admissions for adrenal crisis.

METHODS AND PROCEDURES

Laboratory tests showed adrenocortical hormone (ACTH) 1490 pg/ml and cortisol <1 µg/dl, while a cosyntropin stimulation test was positive. A 24-hour urine metanephrine test was negative. The 17-hydroxyprogesterone (17-OHP) level was 971 ng/dl, likely because the patient was on intermittent steroid replacement, while 21-hydroxylase antibody was normal. Considering the size of the adrenal mass, a biopsy was recommended but the patient refused at that time. Oral steroid replacement was initiated and a follow-up arrangement was made with an endocrinologist, but the patient was lost to follow-up for about a year.

The patient returned to hospital with testicular pain. Examination revealed diffuse skin hyperpigmentation, and bilateral firm testicular enlargement, with the right greater than the left. Laboratory results showed normal beta-human chorionic gonadotropin, alpha fetoprotein

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(AFP) and lactate dehydrogenase (LDH) levels. Testicular ultrasound with heterogeneous enlargement of the testicles bilaterally was noted without identifiable focal lesions as shown in Fig. 2. No hyperaemia was noted. This was most consistent with an infiltrative process with the differential diagnosis including an infiltrative process related to the patient's underlying endocrine abnormalities, amyloid deposition, diffuse enlargement secondary to neoplasm or infectious aetiologies such as orchitis. CT of the abdomen showed similar testicular findings and persistent adrenal glands enlargement. A positron emission tomography scan (PET) showed diffuse uptake in the testicles bilaterally, and hypermetabolic activity in a cervical lymph node and a right inguinal lymph node (Fig. 3), raising suspicion for a primary testicular tumour. Right cervical lymph node biopsy returned negative for malignancy. The patient underwent right orchiectomy due to high suspicion for underlying malignancy. However, the pathology report confirmed a non-neoplastic testicular tumour of adrenogenital syndrome, also known as TART.

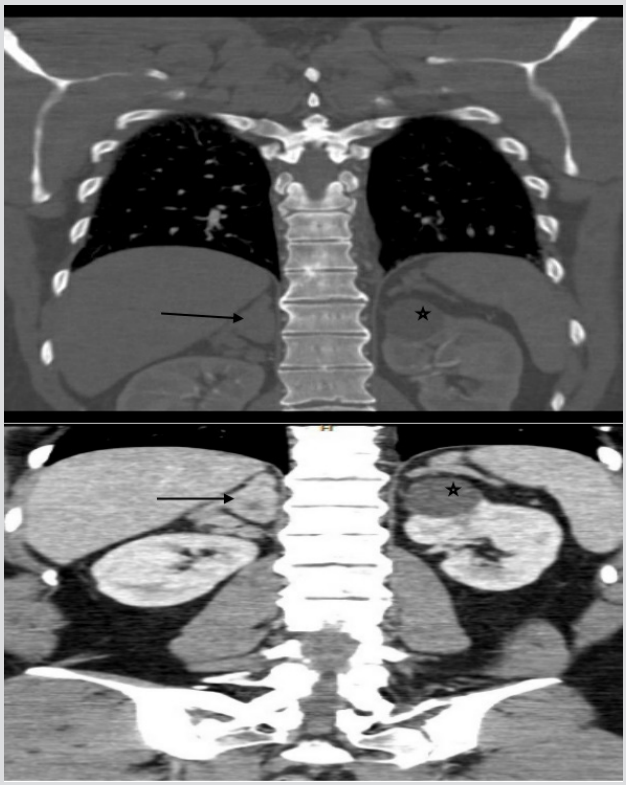

Figure 1. Initial CT of the chest showing incidental findings of enlarged adrenal nodules and a left renal cyst with a left nodular appearing adrenal gland. A CT scan performed on a follow-up visit showed persistent adrenal enlargement. Adrenal enlargement on both images is marked by right arrows and the left renal cyst is marked by star

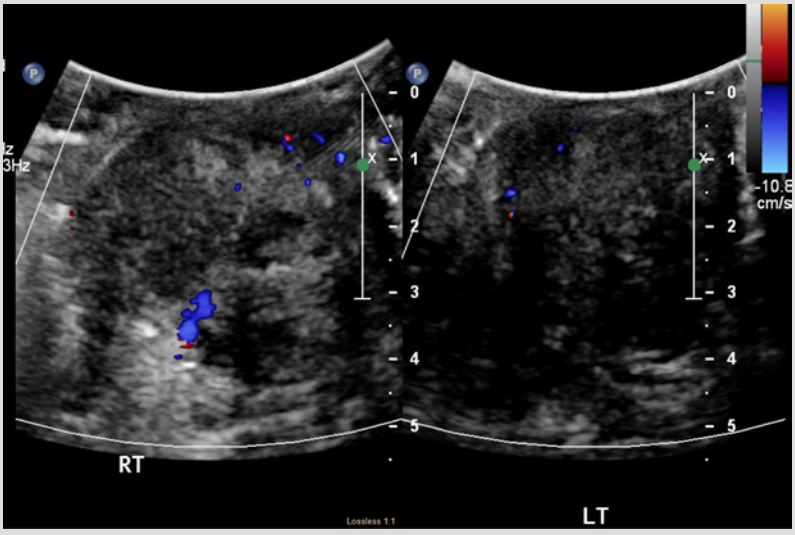

Figure 2. Ultrasound of the testicles showing bilateral heterogeneous enlargement

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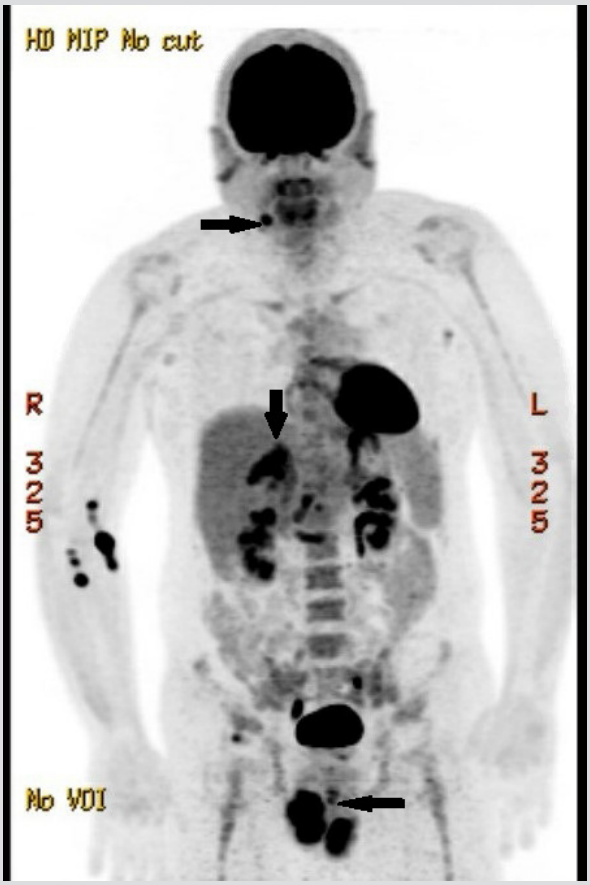

Figure 3. PET scan showing testicular enlargement with lymphadenopathy. Cervical lymphadenopathy is marked by a right arrow, the enlarged adrenal gland by a downward arrow and testicular enlargement by a left arrow

DISCUSSION

Congenital adrenal hyperplasia is characterized by disturbed steroid synthesis, most commonly due to 21-hydroxylase deficiency, leading to high ACTH levels. TART is mainly described in poorly controlled patients but has also been reported in patients with suppressed ACTH and 17-hydroxyprogesterone concentrations. TART is considered to be an aberrant adrenal tissue that proliferates under high adrenocortical stimulating (ACTH) conditions although the exact pathogenesis remains unknown. The reported incidence varies from rare to 94% depending on the methods used [1–3]. Imaging findings are non-specific, while ultrasound usually shows bilateral enlargement of the testes, which are generally located in testicular mediastinum and may appear homogeneous or heterogeneous. In adults, the question of medical versus surgical management arises, especially in the context of anxiety about misdiagnosing malignancy. TARTs may regress with exogenous glucocorticoid and mineralocorticoid replacement, although surgical resection may be needed in larger masses due to compression of normal testicular parenchyma that may lead to infertility [4, 5]. Exogenous steroid replacement can help prevent unnecessary orchiectomy. Imaging findings are non-specific and difficult to differentiate from malignancy. Histologically, TART can appear similar to Leydig cell hyperplasia, causing a diagnostic dilemma in these cases and leading to unnecessary surgical intervention.

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