Case Report

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Congenital Adrenal Hyperplasia and Testicular Adrenal Rest Tumors Causing Infertility and Detected by 18F-FDG PET/CT

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Abstract

Testicular adrenal rest tumors (TARTs) are benign tumors often bilaterally located and are mostly diagnosed in adulthood. TARTs are seen in approximately 94% of patients with congenital adrenal hyperplasia (CAH). In this paper, we present a case of TARTs and CAH detected on 18F-fluorodeoxyglucose (FDG) PET/CT, as a rare case described in only one report in the literature. A 42-year-old patient presented to our outpatient clinic due to testicular masses. The patient had azoospermia, and his ultrasound images revealed bilateral solitary masses. In 18F-FDG PET/CT, high activity uptake was observed in both testicles, and bilateral adrenal hyperplasia was detected incidentally. When the patient's anamnesis was questioned, it was determined that pubarche occurred at the age of nine years, his body weight was 82 kg, and his height was 153 cm. Because of the hormonal evaluation, the patient was diagnosed with CAH and TARTs. In conclusion, 18F-FDG PET/CT may play a decisive role in the evaluation of infertile men with testicular masses.

Keywords: Congenital adrenal hyperplasia, infertility, PET/CT, testicular adrenal rest tumor

Introduction

Testicular adrenal rest tumors (TARTs) are most diagnosed in adulthood and are often located bilaterally (1). In patients with congenital adrenal hyperplasia (CAH), elevated adrenal androgen levels suppress the hypophyseal-gonadal axis, resulting in small testicular sizes and infertility. Obstructive azoospermia and oligospermia caused by TARTs are other causes of infertility (2). Positron emission tomography/computed tomography (PET/CT) performed with 18F-fluorodeoxyglucose (FDG) can help diagnose malignancies in the presence of many suspicious lesions, stage diagnose cases, evaluate treatment response, and detect recurrences with high reliability (3). In this paper, we present a case of CAH and TARTs detected on 18F-FDG PET/CT performed due to testicular masses in a patient who presented to our outpatient clinic with infertility.

Case Report

A 42-year-old patient presented to our outpatient clinic due to testicular masses. The patient had azoospermia according to the sperm analysis. In the blood tests, AFP 1.77 (0.89-8.78 ng/mL) and beta HCG 0.82 (0.22-9.75 mıU/mL) were found. His scrotal Doppler ultrasonography (USG) revealed bilateral vascular solitary lesions containing heterogeneous diffuse calcifications, measuring 32x15x14 mm on the left and 35x15x15 mm on the right, as well as a decrease in both testicular volumes (Figure 1). These lesions were evaluated to be malignant, and 18F-FDG PET/CT was planned. The intense involvement of both adrenal glands was visualized, and bilateral adrenal hyperplasia was detected in the tomography images. The metabolic size of the mass was 9x7 mm maximum standardized uptake value (SUV_{max}): 5.9] in the right testis and 13x12 mm (SUV_{max} 4.3) in the left testis, and high levels of metabolic activities were observed, which were interpreted to correspond to the masses defined on USG (Figures 2-4). These findings were considered to be suspicious in terms of primary testicular malignancy and bilateral adrenal metastases, and the patient was referred to the endocrinology department for re-evaluation for other possible underlying conditions. When the anamnesis of the patient was questioned, it was determined that pubarche had started at the

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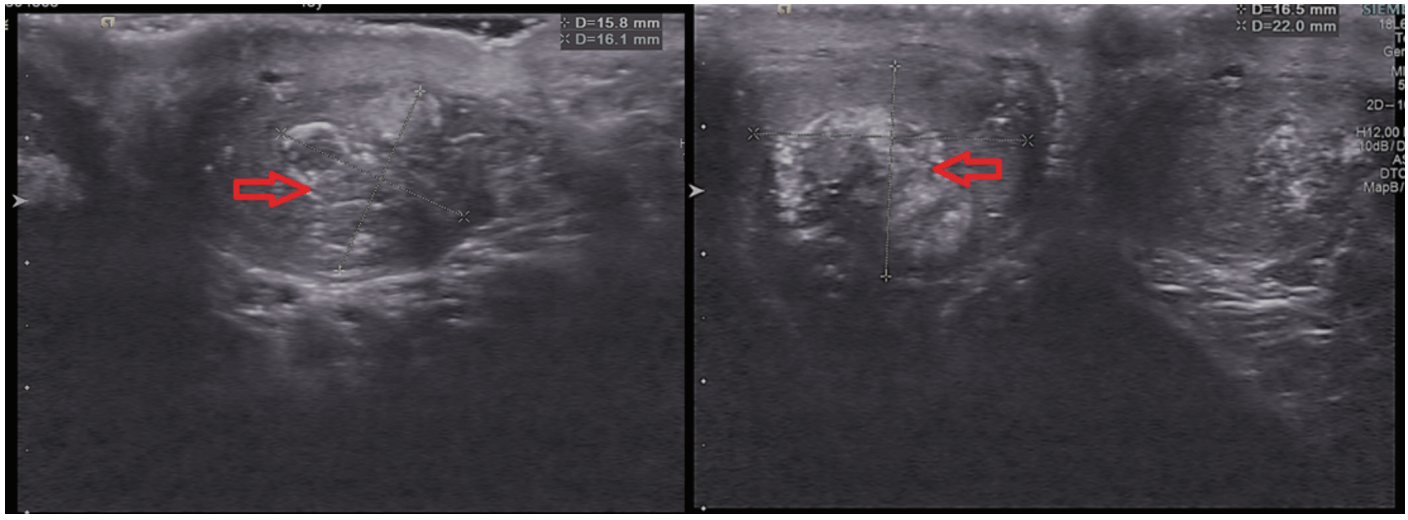

Figure 1. a) Ultrasonography image of the right testicle showing a hypoechoic mass, **b)** Ultrasonography image of the left testicle showing a hypoechoic mass

age of nine years, and his current body weight and height were 82 kg and 153 cm, respectively. System examination findings were normal. The results of the blood tests were as follows: 17-hydroxyprogesterone, >20 ng/mL; androstenedione, >10 ng/mL; ACTH, 306.8 (7.2-63.3) pg/mL; morning basal cortisol, 3.4 (6.2-19.5) mcg/dL; and DHEA-So4, 657 (139-484) mcg/dL. Considering a history of early puberty, short stature, adrenal hyperplasia, bilateral testicular involvement, and elevated androgen levels, a diagnosis of CAH and concomitant TARTs was made, and hydrocortisone treatment was initiated in divided doses of 20 mg/day. Fatigue and erection problems improved after the steroid treatment. In vitro fertilization was planned for infertility treatment. Informed consent was obtained.

Discussion

CAH refers to a group of adrenal steroid synthesis disorders. The disease is autosomal recessive. In these patients, steroid synthesis decreases and consequently the ACTH-level increases. As a result, all cells originating from the adrenal cortex proliferate (1).

TARTs are seen in approximately 94% of patients with CAH (3). These tumors are typically localized in the rete testis and are usually bilateral (4). They are seen in untreated patients and often in young adults (5). One of the most important problems encountered in adult patients with CAH is infertility, as was the case in our patient. In these patients, suppression of the hypophyseal-gonadal axis due to high adrenal androgen levels reduces testicular size and infertility (2).

TARTs are often mistaken for Leydig cell tumors because of their similar features in pathology (6). Microscopic examination of TARTs revealed lobular or nodular cells with extensive eosinophilic granular cytoplasm, separated by fibrous septa. Reinke crystals are characteristic of Leydig cell tumors and seen in 20-40% of cases (4). Leydig cell tumor tends to show a relatively higher metabolic activity on PET/CT (7). However, the diagnosis of TARTs can be made based on a typical history, bilaterality, and characteristic USG and magnetic resonance imaging (MRI) findings. On USG, they are typically seen as bilateral hypoechoic lesions with multifocal acoustic shadowing extending from the hilum to the parenchyma. On MRI, these

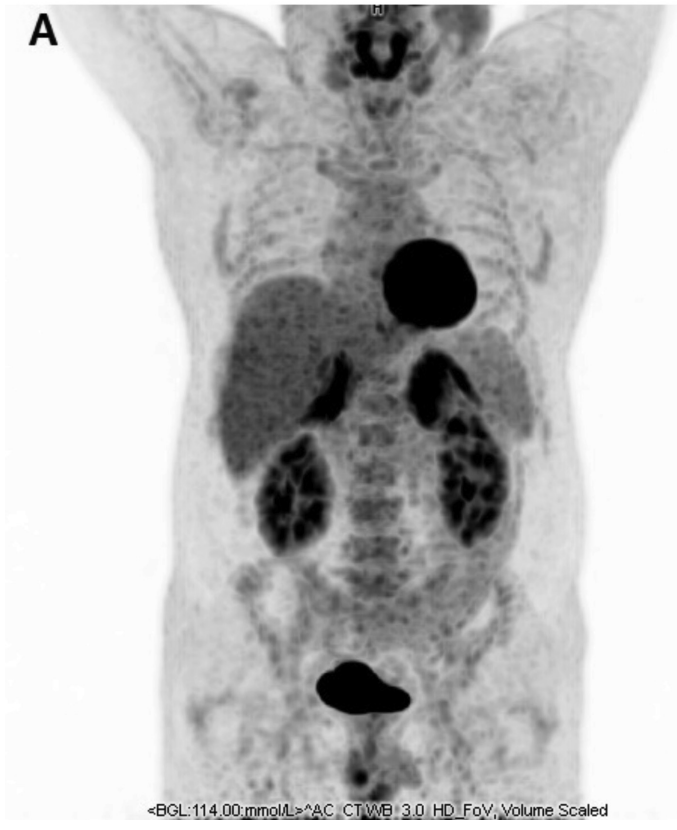

Figure 2. 18F-FDG PET/CT image of congenital adrenal hyperplasia and testicular adrenal rest tumors

FDG: Fluorodeoxyglucose, PET/CT: Positron emission tomography/computed tomography

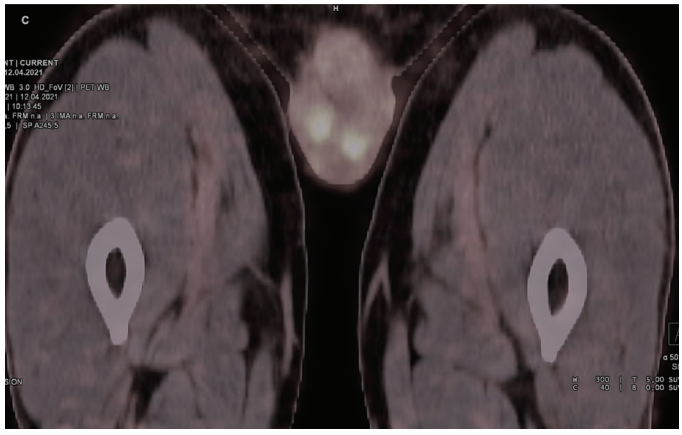

Figure 3. 18F-FDG PET/CT image of testicular adrenal rest tumors

FDG: Fluorodeoxyglucose, PET/CT: Positron emission tomography/computed tomography

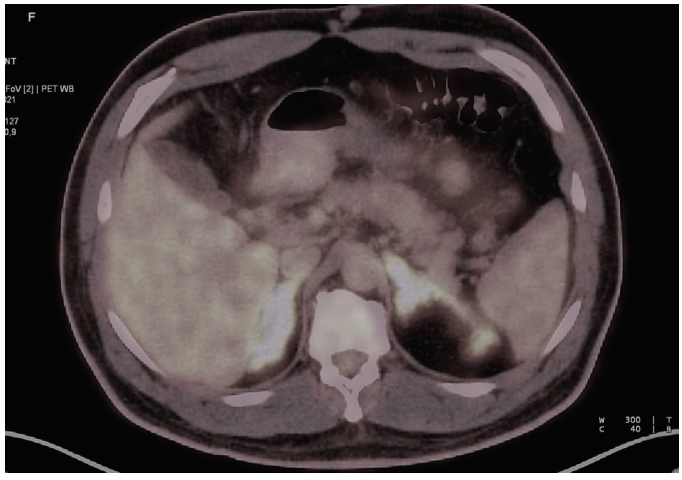

Figure 4. 18F-FDG PET/CT image of congenital adrenal hyperplasia

FDG: Fluorodeoxyglucose, PET/CT: Positron emission tomography/computed tomography

lesions are visualized as bilateral solid masses extending from the hilum to the parenchyma and have contrast enhancement after the contrast agent injection (8). However, it is essential that the endocrine hormone profile supports the diagnosis, and USG and MRI findings alone cannot definitively differentiate TARTs from other testicular malignancies (4).

TARTs may regress in the early period with corticosteroid treatment at a dose that suppresses increased ACTH levels. Response to medical treatment depends on the histological and clinical stage of the tumor (9). In the patient group where medical treatment is not sufficient, if testicular hypofunction has not developed and testicular tissue invasion is not high, testis-sparing surgery can be performed. However, the fertility prognosis in patients with TARTs undergoing testis-sparing surgery remains uncertain. Tumors that cover most of the testicular tissue are considered to be at advanced stages, and orchiectomy is preferred in these patients (10,11).

There is only one case report in which a TART diagnosis was made using 18F-FDG PET/CT in the literature. In this 17-year-old male patient with testicular masses, unilateral orchiectomy was performed, and the pathology result was found to be Leydig cell tumor (12).

Conclusion

CAH and TARTs can be diagnosed following 18F-FDG PET/CT. Our 42-year-old adult patient presented to our outpatient clinic with infertility and was diagnosed with CAH and TARTs based on the 18F-FDG PET/CT findings and endocrine profile before testis loss occurred.

It should be considered that 18F-FDG PET/CT may have a decisive effect in the evaluation of infertile men presenting with a testicular mass.

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Ethics

Informed Consent: Informed consent was obtained.

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