

**Case study**

Prostatic adenocarcinoma in the setting of persistent müllerian duct syndrome: a case report ^{☆, ☆ ☆}



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Summary Persistent müllerian duct syndrome (PMDS) is a form of disordered sex development in which rudimentary müllerian structures are identified in phenotypically and genotypically normal males. It is caused by defects in the anti-müllerian hormone (AMH) system. Since patients with PMDS present with undescended testes, testosterone production by Leydig cells later in life is often decreased. The role of androgens in prostate cancerogenesis is well known. Cryptorchid testes and diminished testosterone levels in post-pubertal life in patients with PMDS play a protective role against prostate cancer, and hence, prostate cancer is a rare event in patients with PMDS. Herein, we present a patient who underwent prostatectomy for high-grade prostatic adenocarcinoma with persistent müllerian structures (such as rudimentary uterus, fallopian tubes, and cervix) identified during surgery. To our knowledge, this is the second case reported in the English language literature where PMDS was associated with prostate cancer.

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1. Introduction

Disorders of sex development (DSD) are medical conditions in which there is disagreement among the chromosomal, gonadal, and phenotypic sex of an individual [1]. Persistent müllerian duct syndrome (PMDS) is a form of DSD in which rudimentary müllerian structures are identified in phenotypically and genotypically normal males. It is caused by defects in the anti-müllerian hormone (AMH) system [2]. Many patients with PMDS present with undescended testes, and due to this, rudimentary müllerian structures are usually incidentally detected during inguinal hernia repair or orchiopexy [3].

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Because most patients with PMDS present with undescended testes, the function of Leydig cells and testosterone production later in life depends on the timeliness of correction of cryptorchidism. Lee et al [4] found an inverse correlation between the age of the surgery (orchiopexy) and testosterone levels and spermatogenesis, revealing a damaging effect when surgery is delayed during childhood.

The role of androgens in prostate cancerogenesis has been widely studied [5,6]. Cryptorchid testes and diminished testosterone levels in post-pubertal life in patients with PMDS play a protective role against prostate cancer. In fact, prostate cancer is rare in patients with PMDS [7]. Herein, we present a patient who underwent prostatectomy for high-grade prostatic adenocarcinoma with persistent müllerian structures, such as rudimentary uterus, fallopian tubes, and cervix, discovered during surgery. To our knowledge, this is the second case in the English language literature where PMDS was associated with prostate cancer.

2. Clinical presentation

In fall 2016, a 64-year-old African-American man presented to the urology clinic with elevated levels (3.1 ng/mL) of prostate-specific antigen (PSA) in the blood. The PSA results measured 10 and 8 years before the current presentation were 0.5 and 0.8 ng/mL, respectively.

His past medical history was noteworthy for osteoporosis, mixed hyperlipidemia, and hypertension, and prostate cancer was significant in his father's medical history. He also reported a history of bilateral inguinal exploration and orchiopexy for bilateral undescended testes when he was 4 years old (in 1956).

The patient reported that he fathered one child, and that he was single (separated from his spouse) at presentation. It is notable that he had been seeing an endocrinologist for male hypogonadism since 2005, and had used Testim (testosterone) gel therapy.

Physical examination showed that the patient had a normal male penis without any deformity. His testes were difficult to palpate but he had scars from inguinal exploration. His prostate was normal in size, without any suspicious nodule or induration.

An ultrasonographical (US) examination performed at presentation showed bilateral inguinal testes. Right and left testes measured $3.6 \times 0.7 \times 2.1$ and $2.8 \times 0.9 \times 2.0$ cm, respectively.

The patient also underwent bone and computed tomography (CT) scans of the abdomen and pelvis. The bone scan showed no osseous metastasis.

Additionally, the CT scan showed a mass-like structure (2.8×3.9 cm), projecting posterolaterally from the base of the prostate on the left side of the pelvis (Fig. 1A).

Although PSA was within the normal range, prostate biopsies were performed because PSA velocity was too high and considered high-risk for cancer. Prostate cancer was diagnosed

on prostate biopsies, and a subsequent robot-assisted radical prostatectomy was performed. No neoadjuvant hormonal treatment was given before the surgery.

3. Materials and methods

The institutional review board was consulted, and no authorization was required for this study. The prostatectomy specimen was processed by the whole-mount method [8]. Briefly, after surgical resection, the specimen was fixed in 10% buffered neutral formalin for 24 hours. Then, the apex and the base of the prostate were amputated, serially sectioned parallel to the urethra, and entirely submitted for further processing. A presumptive seminal vesicle (with adjacent cystic structures) was sectioned perpendicular to the long axis, and representative sections were submitted for further processing. The prostate gland was serially sectioned perpendicular to the urethra at 5-mm intervals and re-fixed in formalin for another 24 hours. From each tissue block, a single section was generated and stained with hematoxylin and eosin. Tumor grade was determined according to the Gleason system.

Additionally, immunohistochemical stains were performed to define the origin of cystic structures adjacent to the seminal vesicle: androgen receptor (AR) (prediluted antibody, 25-min incubation, clone SP107; Ventana, Tucson, AZ), GATA3 (prediluted antibody, 16-min incubation, clone L50/823; Ventana, Tucson, AZ), CD10 (prediluted antibody, 16-min incubation, clone SP67; Ventana, Tucson, AZ), estrogen receptor (ER) (prediluted antibody, 16-min incubation, clone SP1; Ventana, Tucson, AZ), and PAX8 (polyclonal antibody, 1:100 dilution, 15-min incubation; Ventana, Tucson, AZ).

4. Results

4.1. Biopsy findings

A pre-surgical prostate core needle biopsy showed prostate cancer in 3 out of 12 cores of biopsied tissue with Gleason score 9 (4 + 5)-like features in the right mid, right base lateral, and right mid-lateral prostate biopsies, Group V, according to the World Health Organization (WHO), 2016 [9]. There were also 3 other foci showing small atypical glands, suspicious for adenocarcinoma.

4.2. Intraoperative findings

The patient underwent robot-assisted laparoscopic prostatectomy. During the dissection, it was discovered that his right vas deferens crossed over to the left side with both vasa deferentia joining up with what appeared to be a single seminal vesicle. There was no identifiable right seminal vesicle. Also, there was a structure that had the shape of a uterus with a

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