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Serous cystadenocarcinoma of the testis: case report of a rare entity

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Abstract

Background: Germ cell tumors account for more than 95% of testicular cancers followed by sex-cord stromal tumors. The occurrence of serous cystadenocarcinoma in testicular and paratesticular tissue is very rare with distinct clinical behavior.

Case presentation: Herein, we report the case of a 39-year-old man who presented with right testicular swelling. Anatomopathological examination of the right orchiectomy revealed a high-grade paratesticular serous cystadenocarcinoma. Long-term follow-up with close monitoring was adopted in the absence of concomitant metastasis. The patient was disease-free at two years post-surgery.

Conclusion: Owing to its scarcity, there are no standardized guidelines for optimal management of this histological entity. Sharing case reports provide valuable information to support clinical decisions.

Keywords: Serous cystadenocarcinoma, Testicular cancer, Orchiectomy

1 Background

Testicular cancer is the most common solid malignancy in young males, with half of cases occurring between 20 and 30 years old [1]. Germ cell tumors are the first histological diagnosis to consider. Differential diagnosis includes metastasis, ectopic tissue, and ovarian-type epithelial tumors [2]. These rare and unusual histological subtypes should be kept in mind when clinical and paraclinical presentations are suggestive. Most of the reported cases are of serous type, borderline, and occur in middle-age patients. No standard consensus exists regarding the clinical behavior, prognosis and treatment of these neoplasms. The aim of this report is to share knowledge about the management of similar cases that may support clinicians in decision-making from diagnosis to treatment.

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2 Case presentation

A 39-year-old man presented to the surgery department with a 3-month history of progressively growing and painless right testicular mass, without a decline in general condition. The patient reported a 3-year history of infertility after marriage. He had no known medical condition. Physical examination found a nontender scrotal swelling with a negative transillumination test. The contralateral testis was normal and no inguinal adenopathy was observed. Scrotal ultrasound finds a 70-mm solid intratesticular mass that appears homogeneously hypoechoic with no posterior acoustic enhancement and without an image of microlithiasis. Serum level of tumor markers including beta-hCG, AFP, and LDH was within the normal range. CT scan did not show evidence of metastatic disease. The patient underwent a right radical orchiectomy. On gross examination, the orchiectomy specimen was composed of a multilocular half-cystic, half-solid whitish tumor lesion extending over 7.5 cm of long axis (Fig. 1). The histological examination demonstrates a tubulo-papillary tumor proliferation infiltrating the testicular pulp with a morphological appearance suggestive of a paratesticular serous cystadenocarcinoma

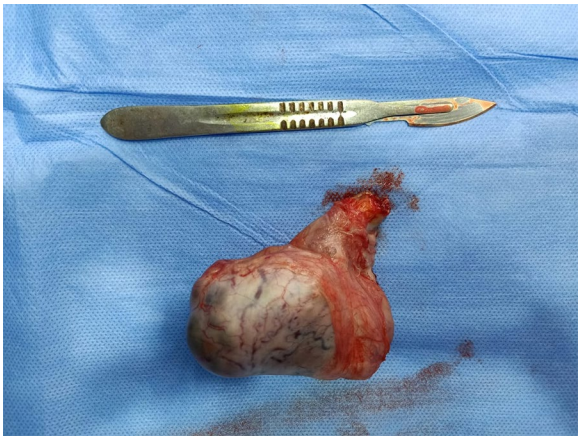

Fig. 1 The orchiectomy specimen on gross examination

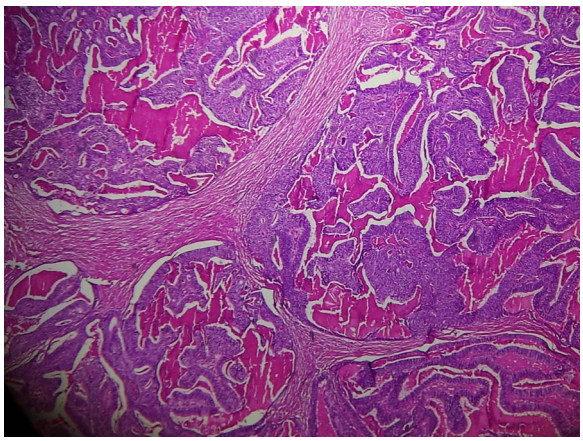

Fig. 3 Tumor proliferation of tubular and papillary architecture

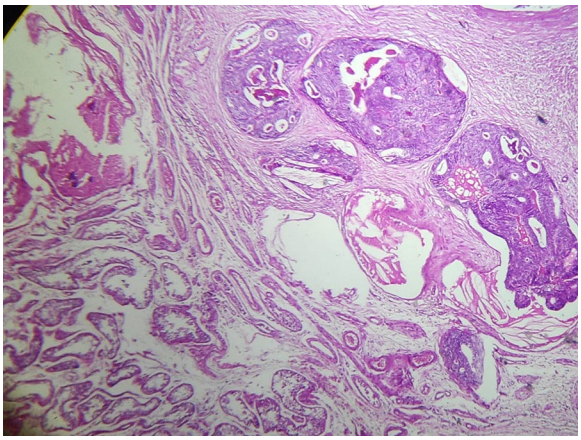

Fig. 2 Testicular parenchyma with cribriform tumor clusters

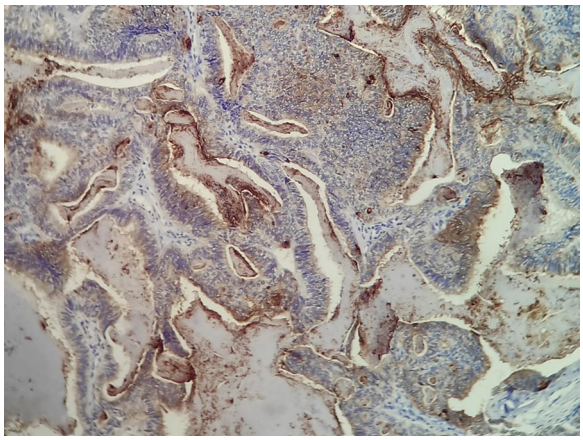

Fig. 4 Diffuse CK7 positive staining in tumor cells

and without evidence of vascular emboli (Figs. 2, 3). The pathological TNM stage was classified as pT1N0M0 S0, stage IA, according to the eighth edition of the AJCC (American Joint Committee on Cancer) cancer staging system for testicular cancer. Immunohistochemical (IHC) staining demonstrated diffuse positivity for CK7 and ACE and negative staining for CK20, CDX2, PSA and WT1. Active surveillance was adopted. The patient's course is uneventful so far, during 2 years of follow-up (Figs. 4, 5)

3 Discussion

Serous cystadenocarcinomas arise most commonly from the ovary epithelium and account for 90% of all ovarian cancers [3]. Often bilateral, both solid and cystic masses, high-grade serous carcinomas have distinctive histologic characteristics based on their etiopathogenesis, typically

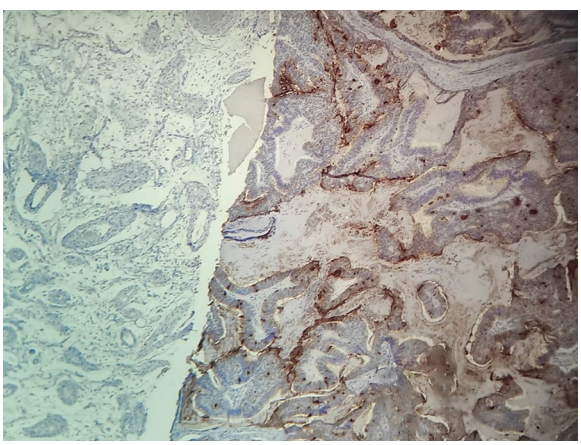

Fig. 5 ACE positive staining in tumor cells

with papillary and glandular growth patterns and high-grade cytologic atypia [4]. The occurrence in the testis or paratestis is very rare and is only reported in a few cases. Testicular serous cystadenocarcinomas share similar morphological and IHC features as their ovarian counterparts and follow the same pathological classification [5].

The clinical presentation is nonspecific with often a history of recent, rapid growth, painless scrotal mass, often found by patient self-exam. Bilateral involvement in men has never been previously reported. Physical examination may reveal inguinal lymph node enlargement or early signs of metastatic disease.

Ultrasound imaging provides a reliable assessment of scrotal masses, by localizing the mass, identifying cysts and cyst-like lesions, and detecting internal blood flow [6]. It allows to eliminate differential diagnosis that can mimic testicular neoplasm (e.g., infections, torsion of the testis, vascular pathologies, or testicular trauma), and evaluate local tumor spread. Serous cystadenocarcinomas are typically heterogeneously hypoechoic compared to normal testicular tissue, often lobulated with cystic components and calcifications [7]. Computed tomography is indicated for disease staging using the American Joint Commission on Cancer (AJCC) tumor (T), nodal (N), and metastasis (M), TNM classification for testicular cancer [8].

These tumors are histologically classified as miscellaneous tumors in the WHO classification of tumors of the urinary system and male genital organs, with most reported cases being borderline tumors [9]. It is suggested that they derive from persistent or incomplete obliteration of the caudal Müllerian duct remnants or from Müllerian metaplasia of the tunica vaginalis which is identical to serous ovarian cancers [10–12]. Elevated serum CA-125 level with normal serum levels of germ cell tumor markers (AFP, BHCG, and LDH) might be helpful to evoke the diagnosis [13, 14].

Radical inguinal orchiectomy establishes the histopathological diagnosis, assesses the local staging of the tumor and is the gold standard treatment with curative intent. On gross description, tumors are variable in size, often a large exophytic cystic mass with papillary growth, and a solid component with necrosis and hemorrhage. Microscopic examination often finds hierarchical glandular patterns, with significant nuclear atypia and pleomorphism, high mitotic index, and high proliferative index -Ki67-. On IHC, tumors cells show positive stains for CK7, PAX8, WT1, p16, and ER and negative stains for CK20 and Napsin A antibodies [2]. P53 gene expression, although rarely performed, appears to be aberrant in almost all cases of high-grade serous cystadenocarcinomas [15], and the $BRAF_{V600E}$ mutation, commonly seen in low-grade ovarian epithelial tumors, is recently reported in a case of a borderline serous tumor of the testis [16].

No consensus has been established about the management of this much rarer entity of serous cancers. Therapeutic approaches are proposed to be similar to those adopted for ovarian serous tumors. Inguinal radical orchiectomy with standard high ligation of the spermatic cord and tumor free-surgical margins is the reference treatment for nonmetastatic testicular serous cancers. Inguinal lymphadenectomy should be performed in patients with macroscopically suspicious lymph node involvement in preoperative imaging [17]. Extemporaneous examination can be performed if the diagnosis of malignancy is doubtful. The role of systematic therapy has not been reached given the known chemoresistance of these tumors. No case reports have previously reported the use of chemotherapy in the adjuvant setting [18]. In metastatic disease, the use of standard platinum-based ovarian chemotherapeutic regimens is indicated but has low likelihood of response as documented in some case reports [19, 20]. Long-term follow-up with close monitoring is required as these tumors have a rapid growth rate and a high risk of distant metastatic relapse. Physical examination, control CT-scan and serum level of CA125 are required for the routine surveillance. However, the optimal follow-up strategy is not clearly determined.

4 Conclusions

Serous cystadenocarcinoma of the testis is an unfamiliar entity to surgeons, pathologists, and oncologists. The probability of its occurrence, although very rare, should be considered in male patients with a suggestive clinical and radiological presentation. Overall, following the same diagnostic and therapeutic procedures adopted for serous ovarian carcinomas is reasonable. In the absence of well-established guidelines and recommendations, sharing case reports is essential to support our clinical decisions.

Abbreviations

BHCG: Beta-human chorionic gonadotropins; AFP: Alpha-fetoprotein; LDH: Lactate dehydrogenase; CK7: Cytokeratin 7; ACE: Angiotensin-converting enzyme; CK20: Cytokeratin 20; CDX2: Caudal-type homeobox transcription factor 2; PSA: Prostate-specific antigen; WT1: Wilms tumor 1.

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Author contributions

ME conceived and designed the manuscript. ME and NB drafted the manuscript. IS and RB critically reviewed, revised, and approved the manuscript. All authors have read and approved the manuscript and agreed to the submission.

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M. El Fadli and N. Benhima have full access to data and are responsible for its integrity. Data is available upon request.

Declarations

Ethics approval and consent to participate
Approval by an ethics committee or institutional review board is not necessary.

Consent for publication
Written informed consent is obtained from the patient for publication of this case report and accompanying images. A copy is available upon request.

Competing interests
Authors declare no conflict of interest.

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