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Characterizing high-grade serous papillary carcinoma of tunica vaginalis

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ABSTRACT

Serous carcinomas of the testis or para-testis are extremely rare tumors of Mullerian type. We report a case of high-grade serous papillary carcinoma of the tunica vaginalis, treated with radical orchiectomy and hemi-scrotectomy after being referred for a rapidly growing painless scrotal mass. In addition to negative testicular tumor markers, scrotal ultrasound, and conventional computerized tomography (CT) scanning, this patient's workup included a positron emission tomography (PET) scan using F-18-fluoro-deoxyglucose (FDG), demonstrating metabolically avid uptake of the disease. This patient is completing ongoing close follow up and is currently disease free at nine months post definitive treatment.

Introduction

Serous carcinoma of the tunica vaginalis (SCTV) is an extremely rare Mullerian-type of tumor, with less than 50 reported cases in the literature. The described diagnosis and management path have typically involved analysis of tumor markers, ultrasound, biopsy, and subsequent staging imaging scans such as computerized tomography (CT). The use of positron emission tomography (PET) imaging with F-18-fluorodeoxyglucose (FDG) radiotracer has not been previously described. In this report, we present a 63-year-old patient with SCTV who underwent surgery with curative intent with close follow-up and surveillance imaging.

Case presentation

A 63-year-old Caucasian male office-worker presented to a tertiary referral centre with a three-month history of a rapidly enlarging painless scrotal mass. He was systemically well on presentation. The patient's medical history included dyslipidemia, hypertension, type-2 diabetes mellitus, and colonic polyps. He is an ex-smoker. He had no previous occupational exposure to asbestos. Clinical examination of the scrotal mass demonstrated a fluctuant unilateral scrotal swelling. No extension or nodal enlargement was palpable in the inguino-femoral region. On referral, his serum alpha-fetoprotein, beta-human chorionic gonadotropin, and lactate dehydrogenase levels were within normal limits. His ultrasound scan demonstrated a large hydrocele with multiple extremely vascular nodular masses arising from the tunica vaginalis (Fig. 1A).

Fine needle aspiration cytology, performed on fluid from the mass, demonstrated an atypical mesothelial proliferation, positive for calretinin and WT1 protein by immunohistochemistry, suspicious for mesothelioma. Staging was performed with both CT and FDG-PET/CT. The diagnostic CT confirmed a large cystic collection with contrast-enhancing solid contents within the left hemi-scrotum, with no nodal or distant spread. The FDG-PET/CT scan demonstrated a localized photopenic mass within the hemi-scrotum with high peripheral metabolic activity corresponding to the areas of contrast-enhancing papillary masses. There was low activity in the central component of the mass, suggestive of necrotic or hemorrhagic regions of disease (Fig. 1B). There was no evidence of primary abdominal mesothelioma or metastatic spread.

The patient was discussed at a uro-oncology multidisciplinary meeting and the decision was made to undergo a hemi-scrotectomy. Adjuvant chemotherapy or radiotherapy was not considered, due to absence of radiological evidence of distant metastatic disease. In concert with the sarcoma surgical team, the patient was consented for a

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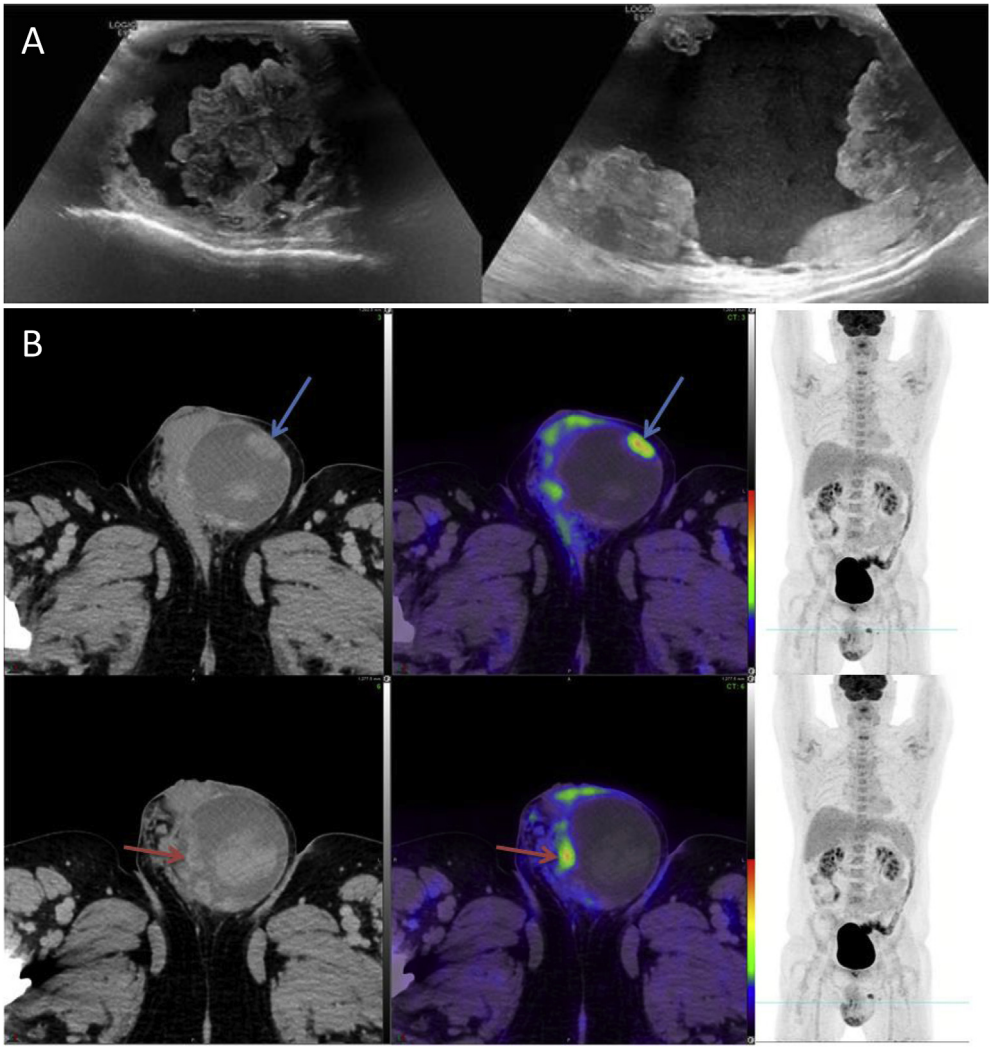

Fig. 1. Ultrasonographic and FDG-PET imaging of the scrotal mass.
The ultrasonographic features of the scrotal mass [Fig. 1A] have been displayed in two images. There is a large hydrocele visualized, with nodular masses arising from the tunica vaginalis. The FDG-PET features of the mass [Fig. 1B] demonstrate a localized photopenic mass within the hemi-scrotum with high peripheral FDG-avid metabolic activity, with corresponding areas identified on CT [*blue & red arrows*].

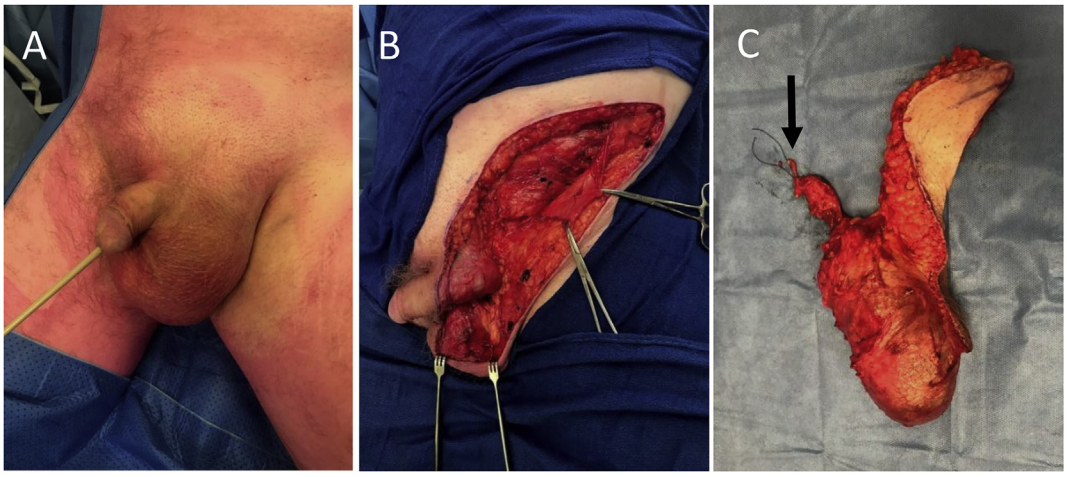

Fig. 2. Intra-operative images of resected specimen and site of resection.
Pre-operative image of the scrotal mass is shown [Fig. 2A]. Intra-operatively [Fig. 2B], artery forceps are reflecting the external oblique aponeurosis and roof of inguinal canal. The resected inguino-scrotal specimen [Fig. 2C] is demonstrated, with a marking suture [*black arrow*] indicating ligation of the cord at the level of the deep inguinal ring.

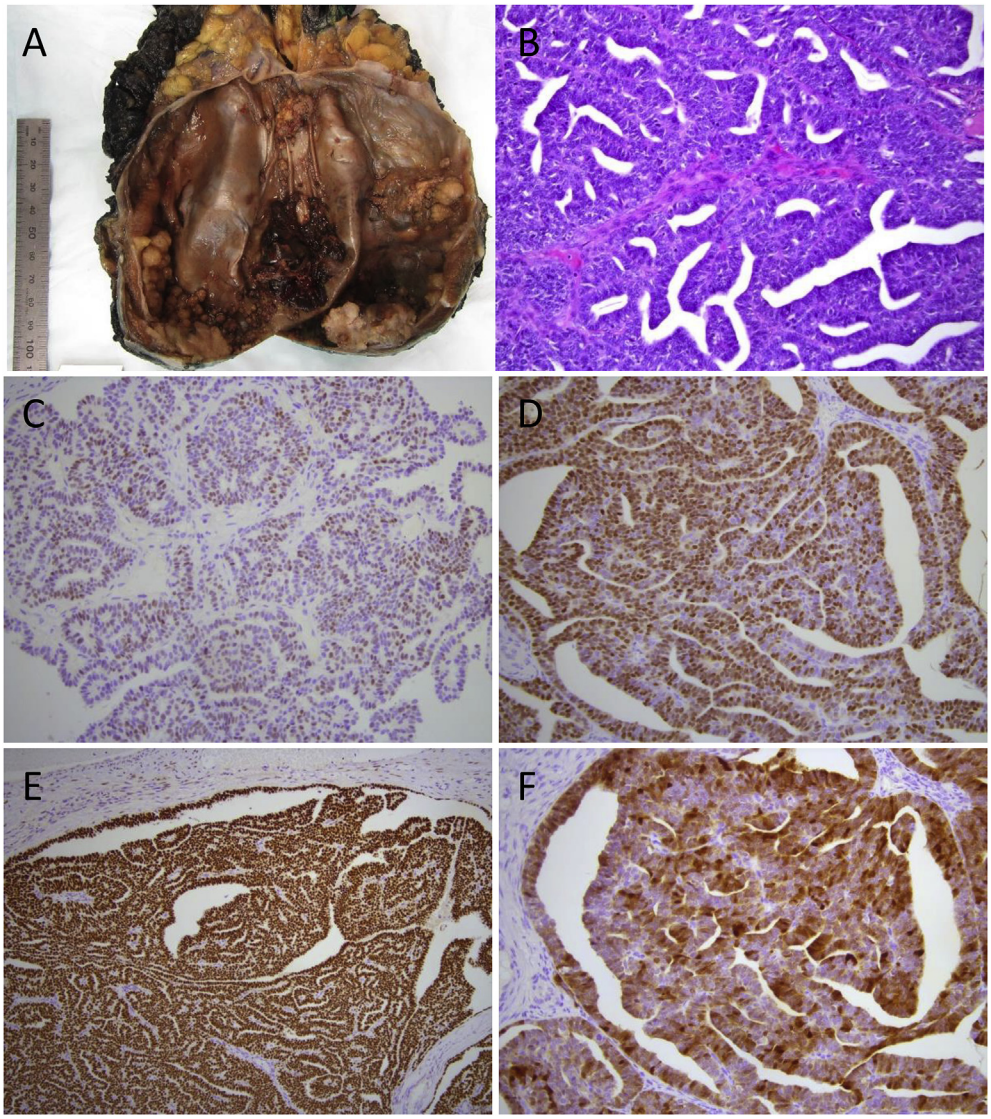

Fig. 3. Macroscopic and microscopic images of the resected specimen.
The resected mass demonstrates an 813g 276 × 75 × 75mm inguino-scrotal resection, with a 144 × 75 × 75mm cystic mass filled with loose hemorrhage [Fig. 3A]. Hematoxylin and eosin-stained section showing irregular slit-like glandular spaces, formed by markedly atypical cells (original magnification x200) [Fig. 3B]. Immunohistochemistry shows reactivity of tumor cells for oestrogen receptor ([Fig. 3C], original magnification x200), progesterone receptor ([Fig. 3D], original magnification x200), WT1 ([Fig. 3E], original magnification x100) and calretinin ([Fig. 3F], original magnification x200).

radical orchiectomy plus left hemi-scrotectomy plus potential groin dissection (Fig. 2).

Subsequent tissue pathology revealed a diagnosis of high-grade SCTV with focal invasion. Macroscopic examination of the resected mass demonstrated an 813 gm 276 × 75 × 75mm inguino-scrotal resection, with a 144 × 75 × 75mm cystic mass filled with loose hemorrhage (Fig. 3A). Multifocal tumors, the largest measuring 75 × 58 × 21mm, arose from the tunica vaginalis with a friable papillary appearance. The testis, measuring 30 × 20 × 11mm, was compressed along the posterior wall, along with the epididymis, rete testis and spermatic cord. The biopsy tract was excised *en bloc* with the specimen. On microscopic examination, the tumors were composed of atypical epithelioid cells forming irregular, slit-like glands and papillae covered by multi-layered epithelium, with tumor cells possessing large, crowded ovoid nuclei with prominent nucleoli and moderate amounts of eosinophilic cytoplasm (Fig. 3B). There were frequent mitotic figures, and large areas of necrosis. Focal invasion was apparent in underlying fibroadipose tissue. No lymphovascular space invasion or perineural permeation was seen. Clear radial surgical margins were obtained. Immunohistochemistry showed that tumor cells were diffusely positive for cytokeratin AE1/AE3, WT-1 (nuclear), calretinin, estrogen receptor, progesterone receptor, cytokeratin 7 and Ber-EP4. BAP-1 expression was retained, and p53 showed wild-type expression (Fig. 3C–F). The tumor cells were negative for thrombomodulin, caldesmon, cytokeratin 5/6, D2-40, Pax-8, EMA, cytokeratin 20, TTF-1, CDX-2, GATA-3, and inhibin.

Currently, this patient has been followed up post-operatively for nine months. The patient was reviewed three months post-operation with CT imaging, demonstrating no clinical or radiological residual or recurrent disease. A subsequent review was made six months after the first review, again with CT imaging. The patient was negative for any recurrence at this review. The patient is planned for ongoing six-monthly reviews with surveillance imaging.

Discussion

High-grade SCTV is an extremely rare disease. Most cases reported have been in patients typically describing a rapidly developing painless scrotal mass, with or without an associated hydrocele.[1] The initial imaging investigation has been commonly performed with ultrasonography. However, ultrasonographic features can be equivocal and challenging to interpret.

The use of FDG-PET imaging demonstrated in our case suggests utility for characterizing the tumor as being metabolically avid. This will be useful in detecting and assessing nodal or distant metastatic disease, in both the initial staging and post-operative surveillance period.

Although previous reports have been published in the field of serous

carcinomas, our report addresses the management of specifically a high-grade disease arising from the tunica vaginalis.[2,3] In addition, to further assess the disease before metastatic spread, the use of PET imaging with FDG radiotracer has been newly described in this case.

The suspected diagnosis of malignant mesothelioma obtained via fine needle aspiration cytology reflects a previously described overlap with serous carcinomas, particularly in the context of a limited sample. Upon review of literature, a limited diagnostic panel helpful in differentiating the two tumors include BER-EP4, B72.3, BG8, calretinin and desmin – desmin is useful for excluding reactive mesothelial proliferation in contrast to malignant mesothelioma.[4] In the resection specimen, the tumor morphology, in conjunction with the expression of BER-EP4 and estrogen and progesterone receptors, and absence of expression of cytokeratin 5/6, thrombomodulin and podoplanin, lead us to render a diagnosis of SCTV.

Primary treatment of high-grade SCTV has been previously suggested as radical inguinal orchiectomy with hemi-scrotectomy. In previous literature, adjuvant chemotherapy has also been offered as per treatment for ovarian serous carcinomas, however a previous report suggests there may be resistance to these agents.[5] Despite this, the management of this extremely rare tumor should consist of early referral, early diagnosis, and minimal delay to primary treatment. The use of FDG-PET imaging will benefit in the staging and ongoing surveillance of disease.

Conclusion

SCTV is a rare tumor that can be mistaken for malignant mesothelioma on radiological or pathological examination. PET imaging may also be considered as an additional useful modality for initial staging and monitoring of recurrence.

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