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Epithelioid sarcoma of the penis: A penile sparing approach, and long-term implications

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ABSTRACT

A 33-year-old male presented with a one-centimetre lesion at the penoscrotal junction which was excised and revealed to be an epithelioid sarcoma (ES). A wide local excision of the lesion and subsequent neoadjuvant radiotherapy followed, with transposition of the patient's testicles laterally to protect fertility. At 3-year follow-up, the patient has no local or distant recurrence but does have a low sperm count. The patient has also had intermittent haematospermia since his treatment for which a cause has yet to be identified. This case highlights that ES of the penis can be managed successfully with surgical excision and local radiotherapy.

1. Introduction

Epithelioid Sarcoma (ES) is a rare malignancy with high rates of local recurrence and metastatic disease. This case discusses a rare occurrence on the penoscrotal junction and subsequent management.

2. Case presentation

A 33-year-old Caucasian male presented with a nodular one-centimetre non-tender subcutaneous lesion at the penoscrotal junction, concerning for malignancy. He was previously well with no significant background history and a non smoker. Positron Emission Tomography (PET) imaging showed no regional or metastatic disease.

The patient underwent a cystoscopy and excisional biopsy. His cystourethroscopy showed 5 mm of mild concentric circular narrowing in the anterior urethra adjacent to the lesion. The cystoscopy was otherwise unremarkable. On excision, the penile lesion was found to invade the corpus spongiosum without breaching the urethra. Frozen section of the margin showed no signs of malignancy. A dartos flap was mobilised and used as a graft to repair the defect in corpus spongiosum. The patient recovered well, had a successful trial of void and was discharged home.

Histologically, the tumour was an Epithelioid Sarcoma with a multilobulated mass, infiltrative edge and central coagulative necrosis. cytologically it was composed of epithelioid cells with pelomorphic rhomboid nuclei, pale eosinophillic cytoplasm and ill-defined borders. Immunochemically, it was positive for cytokeratin AE1/AE3, EMA, D2-40 and ERG. There was no staining with CD34, CD32 or CK5/6 (Fig. 1). A wide local excision was subsequently performed two months after the excisional biopsy, requiring further excision of the corpus spongiosum and urethroplasty with buccal mucosal grafting. This second specimen showed no evidence of malignancy. The patient was discharged home without complication.

Adjuvant local radiotherapy was recommended at the uro-oncology multidisciplinary team meeting, but the patient was quite concerned about infertility with this treatment. Pre-treatment, the patient had normal FSH, LH and testosterone hormone levels, and semen analysis showed normal sperm concentration, morphology and motility. Three months post his initial excisional biopsy, bilateral transposition of the testicles lateral to the external inguinal ring was performed prior to his radiotherapy. A total of 64Gy was delivered in 32 fractions as adjuvant therapy for local control. Post-radiotherapy, an orchidopexy was performed to relocate his abdominal testes into his scrotum.

At 3-year follow up, PET confirmed no local or distant malignancy. The patients' partner conceived using sperm stored prior to radiotherapy as our patient's sperm count and concentration were low. Unfortunately, on recent semen analysis, these have not returned to baseline but his LH, FSH and testosterone are at normal levels. The patient described occasional haematospermia associated with erections in which he passes large clots. This was investigated with Transrectal Ultrasound (TRUS), Magnetic Resonance Imaging (MRI), PET and pelvic

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angiography however, showed no cause for haematospermia. A urethroscopy was performed during a medically induced erection revealing a bleeding point from the graft site, which was found and cauterised. Since the procedure, his haematospermia has improved, but still persists.

3. Discussion

Penile malignancy is rare, accounting for 0.5 % of all malignancies.[1] Around 95 % of these malignancies are Squamous Cell Carcinoma with the remaining 5 % comprising of Basal Cell Carcinoma, ES, amongst others.

ES is a rare soft tissue sarcoma typically occurring in the distal extremities of young adults.[1] It classically presents as a slow growing nodule within the deep subcutaneous tissue. ES of the peno-scrotal region accounts for approximately 5–6% of all ES, often presenting as a slow growing, non-tender nodule, but can also present as multifocal disease.[1,2] Historically, management consists of total or subtotal penectomy.[2] Obtaining negative margins is imperative as ES has a strong propensity for local recurrence, as seen in approximately 85 % of cases, with a risk of distant metastasis in 30–50 % of cases.[2] While the efficacy of adjuvant radiotherapy has been proven to improve survival in most soft tissue sarcomas, radiotherapy's efficacy in ES is uncertain due to limited sample sizes and follow up.[2] Despite ES being considered a radiosensitive tumour, it has a poor overall survival. Adjuvant radiotherapy can decrease rates of local recurrence in combination with surgery, yet disease free survival sits at 70 % and 55 % at 5 and 10-year follow up respectively.[2] Our case was managed with wide local excision and adjuvant radiotherapy, and at three years post treatment is free from recurrence.

Radiotherapy of the male genitalia often has a detrimental impact on fertility. Constant production and mitotic activity of spermatogonia make it extremely vulnerable to radiotherapy.[3] Dose rates of less than 1 Gy have been demonstrated to reduce the number of spermatogonia, with long term damage and decrease in sperm count occurring from around 4 Gy.[3] Testicular transposition into the abdominal cavity enables local radiotherapy to proceed without risking infertility.[4] Most studies reporting on fertility rates post radioprotective testicular transposition have focused on children and adolescents with bladder/genital malignancies. To our knowledge, only one case series with two cases has reported the fertility rates of adult patients post testicular transposition.[4] Both patients in this series had normal semen analysis at three months followup post radiotherapy. Our case shows that radioprotective testicular transposition is a potential option that may help preserve adult fertility.

Our patient also experienced haematospermia. He described pain and the passing of large clots with erections. In a patient who has had multiple procedures like ours, the provisional diagnosis of a veno-urethral fistula was made and subsequently excluded through multiple routes of imaging, specifically pelvic angiography. Haematospermia is renowned for its numerous aetiologies including infection, malignancy, trauma and iatrogenic causes.[5] Yet, in around 70 % of cases, the cause of haematospermia is unknown.[5] Investigations for haematospermia differ depending upon history and physical examination. A thorough clinical assessment including urine analysis, full blood count, coagulation profile and prostate specific antigen should be performed.[5] Various imaging techniques may also help determine the aetiology of haematospermia. TRUS, MRI, CT and pelvic angiography are all useful in evaluating haematospermia. However, in our case all aforementioned imaging techniques were performed, and all were non diagnostic, specifically angiography which showed no signs of any fistulae or arteriovenous malformations.

4. Conclusion

This case highlights that peno-scrotal ES can be successfully managed with surgical excision and local radiotherapy and bilateral testicular transposition is a viable option to protect fertility. As with all surgical procedures, complications can occur and haematospermia, if encountered, needs to have a thorough clinical assessment to determine its cause.

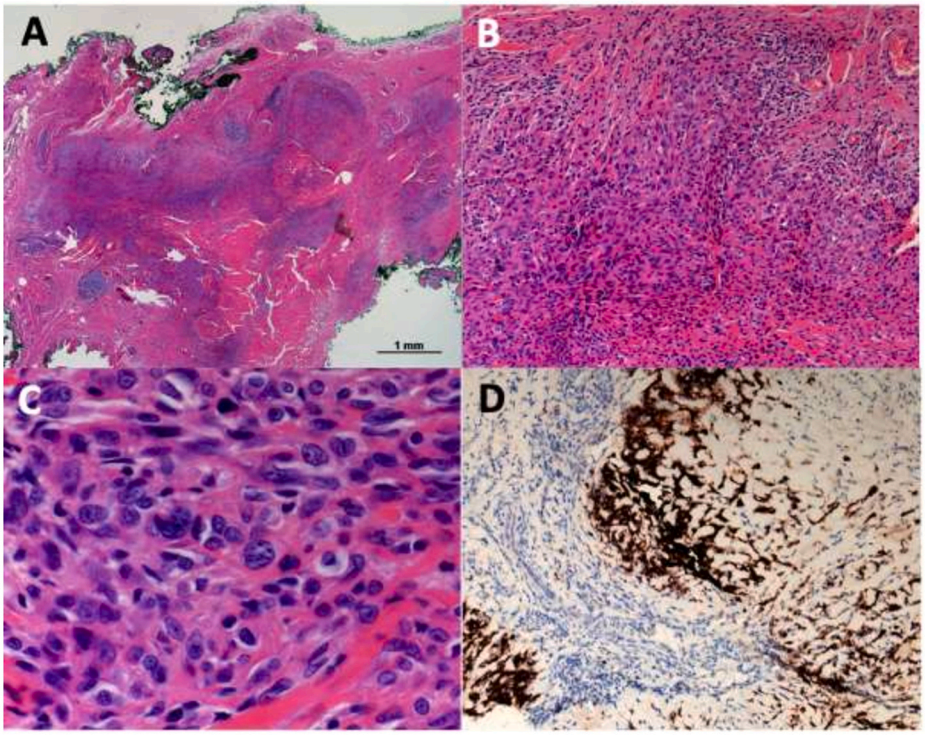

Fig. 1. Histopathology slides showing Epithelioid Sarcoma. Haematoxylin and Eosin Stain at low (A), medium (B), high power (C) and Cytokeratin AE1/AE3 (D).

Declaration of competing interest

None declared.

References

1. Chbani L, Guillou L, Terrier P, et al. Epithelioid sarcoma: a clinicopathologic and immunohistochemical analysis of 106 cases from the French sarcoma group. *Am J Clin Pathol.* 2009;131(2):222–227.
2. Sobanko JF, Meijer L, Nigra TP. Epithelioid sarcoma: a review and update. *J Clin Aesthet Dermatol.* 2009;2(5):49–54.
3. Biedka M, Kuźba-Kryszak T, Nowikiewicz T, Żyromska A. Fertility impairment in radiotherapy. *Contemp Oncol.* 2016;20:199–204.
4. Deo SV, Asthana S, Shukla NK, Mohanti BK. Fertility preserving testicular transposition in patients undergoing inguino pelvic irradiation. *J Surg Oncol.* 2001;76(1):70–72.
5. Mathers MJ, Degener S, Sperling H, Roth S. Haematospermia – a symptom with many possible causes. *Deutsches Arzteblatt Int.* 2017;114(11):186–191.