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Bilateral Contiguous Scrotal Extramammary Paget's Disease Treated With Mohs Micrographic Surgery and CK7 Immunohistochemical Staining

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INTRODUCTION

Extramammary Paget's Disease (EMPD) is a rare malignancy that commonly presents on apocrine-rich skin as a unilateral scaly patch or plaque that often grows slowly and asymmetrically.[1] There is growing evidence to suggest that Mohs micrographic surgery (MMS) may provide better patient outcomes and lower recurrence rates compared to wide local excision. We present a rare case of EMPD presenting as bilateral scrotal patches that were found via cytokeratin-7 (CK7) staining during MMS to connect subclinically in a contiguous fashion around the base of the penis.

CASE REPORT

A 60-year-old man from South Korea was referred for MMS with CK7 for biopsy-proven EMPD on the right and left scrotum. He was scheduled for wide local excision in South Korea prior to referral. His physician learned about the Mohs technique and the improved cure rates and made arrangements for referral and transfer of care. Physical examination revealed a 5.5 cm x 5 cm pink patch with features of EMPD on the left scrotum (Figure 1), and a 7 cm x 5 cm subtle pink and dyspigmented patch on the right scrotum (Figure 2).

FIGURE 1. (A) Pink scaly patch with features of EMPD on the left scrotum. (B) Subtle pink and dyspigmented patch on the right scrotum.

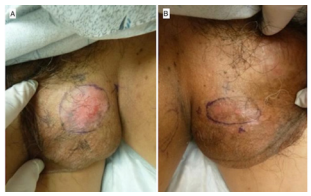
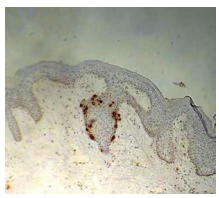


FIGURE 2. CK7 immunostaining of scouting biopsies beyond the visible margins of the tumor was consistent with EMPD, highlighting individual Paget cells confined to the epidermis.



During the preoperative consultation, scouting biopsies with CK7 staining were performed on each side, at 12, 3, 6, and 9 o'clock positions beyond the visible margins of the tumor to better assess the extension of EMPD (Figure 3). On the day of surgery, peripheral Mohs technique was used to remove a ring of tissue beyond the visible border and the positive scouting biopsies.[2] MMS using CK7 staining was performed to improve accuracy of margin control, as highlighted in several recent studies.[2-4]

FIGURE 3. Five stages of MMS revealed that the defects on the left and right scrotum were essentially contiguous in the suprapubic region and perineum. The defect was repaired with a large, meshed skin graft.

MMS required two days and five stages at which point the defects on the left and right scrotum were essentially contiguous in the suprapubic region and perineum. A small finger-like projection of EMPD was still positive in the inferior perineum. The patient refused to proceed with additional stages of Mohs in the office. The collaborating plastic surgeon removed the islands of tumor left in the center of the wound after peripheral margins were cleared. He also excised the positive area in the inferior perineum; en face processing with CK7 revealed cleared margins. The large contiguous defect on the right and left scrotum, which collided in the suprapubic region and below the scrotum, was repaired with a large, meshed skin graft (Figure 3).

DISCUSSION

EMPD often presents similar to an inflammatory skin condition, resulting in misdiagnosis and delayed treatment. Primary EMPD arises as an intraepidermal neoplasm of glandular origin.

A simplified classification of EMPD has been proposed and the authors are in agreement with this classification: Type I EMPD is defined by Paget cells confined entirely to the epidermis and the epithelial structure of its adnexa, with or without dermal invasion. Type II EMPD exists within the epidermis, the epithelial structure of its adnexa, and the contiguous epithelium of the genitourinary or gastrointestinal tract.[5] Many of the underlying cancers seen in EMPD patients are likely coincidental and unrelated to the EMPD or represent contiguous disease extending into the lower gastrointestinal or genitourinary tract (Type II).[5] The search for underlying cancers in EMPD patients should be focused on contiguous disease extending into the urethra, anal canal, and vaginal/cervical mucosa if there is clinical suspicion or if indicated by the scouting biopsies or surgical margins. Regardless of EMPD, all patients should have age appropriate screening for other cancers per recommended guidelines.

EMPD predominantly affects females, though the male:female ratio nears 1:1 amongst Asian patients.[3] Involvement of the male genitalia comprises only 14% of EMPD cases; these lesions are usually unilateral and appear on the scrotum, penis, groin, and perianal regions.[1] Bilateral involvement of the male genitalia is even more rare, and there have been few case reports of bilateral EMPD involving the genitalia and/or axilla.[6,7] Thus, our patient presents a rare case of EMPD with bilateral involvement of the scrotum that connected subclinically in a contiguous fashion, as revealed by MMS with CK7 staining.

Surgical removal is the mainstay of treatment for EMPD; however, wide local excision results in high recurrence rates, ranging from 22% to 60%, which may be due to the wide subclinical spread of EMPD outside of empirical surgical margins.[8,9] There is growing evidence that MMS results in more favorable patient outcomes, with recurrence rates ranging from 8% to 26% (16% in primary EMPD, 50% in recurrent EMPD), with an overall cure rate of 100%.[8,9] Though MMS allows for 100% margin evaluation and more complete tumor removal than wide local excision, recurrence rates for EMPD treated with MMS remain higher than for other cutaneous malignancies, such as basal cell and squamous cell carcinomas (3.5% recurrence), treated with MMS.[10] The relatively high recurrence of EMPD after MMS is proposed to be due to the difficulty in recognizing single tumor cells on routine staining.[3] Thorough identification of tumor cells is especially important for EMPD lesions, which may show a highly irregular, subclinical growth pattern with finger-like projections that do not stain with hematoxylin & eosin, yet stain with CK7.[2] In a retrospective, multi-center, cross-sectional study of patients treated with MMS with CK7 staining, all primary and recurrent EMPD tumors that were excised entirely by MMS and fully evaluated with intraoperative CK7 immunostaining had a composite local recurrence rate of 3.3%, similar to that of basal cell and squamous cell carcinoma treated with MMS.[3]

In this patient, standard surgical excision of the two separate lesions based on clinical exam alone would likely have resulted in incomplete removal. Our experience adds to a growing body of literature demonstrating that MMS with CK7 staining results in the most complete removal of EMPD and provides for the lowest risk of recurrence.[3,9] In addition, it supports the previous finding that the tumor extends far beyond the visible margins in a finger-like contiguous pattern.[2] Preoperative scouting biopsies with CK7 staining is strongly advised in planning surgical excision.

DISCLOSURES

The authors have no relevant conflicts.

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