

Management of extramammary Paget's disease with a staged, modified Mohs technique

Anant Shukla, MD,¹ Matthew Stringer, DO,² Pansy Uberoi, MD,²
Steven Hudak, MD,² George Kallingal, MD²

¹Department of Urology, Madigan Army Medical Center, Tacoma, Washington, USA

²Department of Urology, San Antonio Military Medical Center, San Antonio, Texas, USA

SHUKLA A, STRINGER M, UBEROI P, HUDAK S, KALLINGAL G. Management of extramammary Paget's disease with a staged, modified Mohs technique. *Can J Urol* 2016;23(4):8382-8384.

Extramammary Paget's disease (EMPD) is a rare intraepithelial adenocarcinoma arising from apocrine glands. We describe an innovative surgical technique to manage extensive cutaneous malignancies in a notoriously challenging location. The patient is a 78-year-old male who presented with "jock itch" on his left groin refractory to topical treatment. A shave biopsy of the lesion

demonstrated non-invasive EMPD which yielded a urology consult. Rather than the standard wide local excision (WLE), which can lead to positive margins, progression, and recurrence, we used a modified the Mohs technique and staged the procedure, providing excellent oncologic and cosmetic outcomes. The described technique has particular merit with uncertain margin status and when geography of lesions preclude a standard Mohs surgery.

Key Words: extramammary Paget's disease (EMPD), eczematous lesions, intraepithelial malignancy, Mohs surgery, staged surgical technique

Introduction

Penoscrotal extramammary Paget's Disease (EMPD) was first reported in 1889 by Crocker et al.¹ The estimated incidence of penoscrotal EMPD is 1/3.7 million and it occurs most commonly between the ages of 50-80.² Because of its rarity, penoscrotal EMPD is frequently misdiagnosed as eczema, tinea cruris, or other benign dermatological conditions. Delay in diagnosis is a major risk factor for disease progression.³⁻⁵ Primary penoscrotal EMPD is typically limited to the epithelium and may become invasive but it is rarely associated with a distant adenocarcinoma. In contrast, secondary EMPD is associated with epidermotropic spread of malignant cells from an underlying adenocarcinoma.⁶ Histologically, the two varieties are often indistinguishable; as a result any

patient with EMPD should be screened for a primary occult adenocarcinoma. We recommend cystoscopy, rectal exam, PSA, colonoscopy and axial imaging as clinically indicated.

Wide local incision (WLE) has traditionally been the mainstay of treatment for EMPD but recurrence is common, due in part to the inherent difficulty in obtaining negative margins.^{4,5} There are numerous case reports demonstrating non-congruence between frozen section and final permanent pathology results. The urologist must be prepared for interval re-resection before performing a complicated wound closure. To that end, we present a modification of WLE and traditional Mohs surgery which allowed us to determine accurate margin status before undergoing a definitive excision and closure.

Case presentation

A 78-year old Caucasian male with a healthy active lifestyle, presented to his primary care provider with a left groin "jock itch." After approximately 2 years of treating the lesion with topical steroids, antifungals and

Accepted for publication July 2016

Address correspondence to Dr. Anant Shukla, Department of Urology, Madigan Army Medical Center, 9040 Jackson Avenue Joint Base Lewis-McCord, Tacoma WA 98431 USA

bacitracin ointment the primary care provider referred the patient to a dermatologist who performed a shave biopsy confirming the diagnosis of EMPD, which based upon the location of the lesion, led to a urology referral. On physical exam the patient had a pruritic, eczematous 8 cm x 8 cm x 8cm lesion with lichenification of the left groin, irregular borders and no skip lesions. The focus of the lesion was in the left groin intertriginous fold but it extended to the lateral scrotum, base of the penis and the medial thighs. The patient's medical history was significant for low risk prostate cancer, status post radical prostatectomy in 2002, without evidence of recurrence. He had also undergone WLE of a melanoma on his shoulder with negative margins. The malignancy was staged with clinic cystoscopy, colonoscopy, PSA/DRE and contrasted CT scan of the abdomen and pelvis. No additional malignancies or lymphadenopathy were noted.

Surgical technique/management

We placed the patient in the lithotomy position and, under general anesthesia, obtained a 5 mm punch biopsy from the center of the lesion as a histologic reference point. Then, using a clock-face pattern we then sent



Figure 1. Modified Mohs technique with "clock-face" punch biopsies demarcating the grossly visible area of the lesion.



Figure 2. Wide local excision of the lesion following demarcation of the malignant area via the modified Mohs technique.

additional punch biopsies in 30 "minute" increments to demarcate the boundaries of the specimen 1 cm away from visibly abnormal skin, Figure 1. With seamless communication between the surgical team and the pathologist, we delivered the multitude of carefully labeled specimens for frozen section. While awaiting the frozen results we excised the intervening skin, Figure 2. Two of the punch biopsies from the left groin were suspicious for EMPD so an additional 1 cm margin was taken at these locations in stage one. Rather than close the wound at this point, a Wound Vac device was placed while final margin status was confirmed. Total operative time was 2.5 hours which included time spent communicating face to face with the pathologist. The final margins from stage one were positive in two non-contiguous regions and so 2 weeks after stage 1, the two areas of interest were re-resected with an additional 1 cm margin and the wound was closed with a rotated rectus abdominis muscle flap, Figure 3. Total operative time for stage 2 was 3 hours. No postoperative complications were encountered after stage 1 or stage 2. At a 5 month follow up the patient was Eastern Cooperative Oncology Group (ECOG) 0, described no voiding difficulties had negative final margins and excellent cosmetic results.

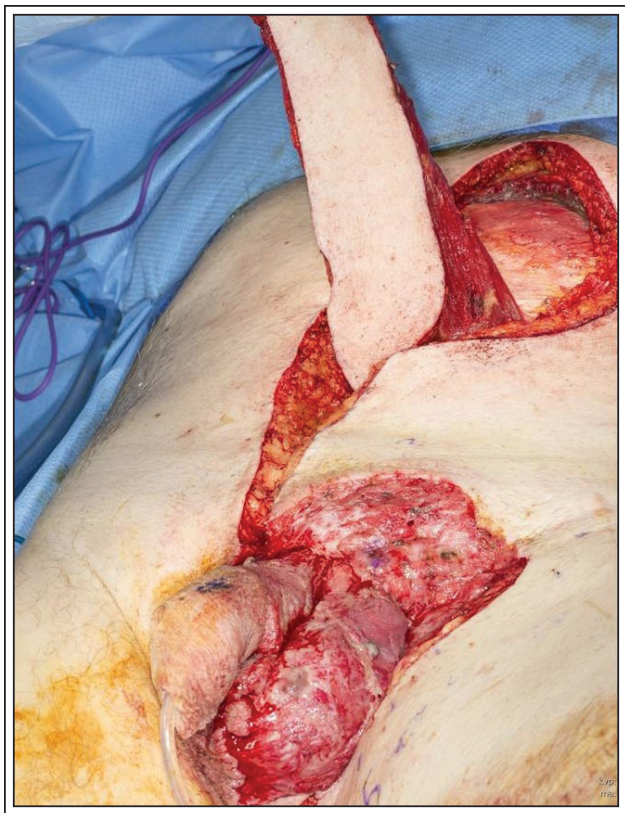


Figure 3. Harvesting of rectus abdominal flap for closure of the defect, following confirmation of negative margins by pathology.

Discussion

While some authors have described topical, systemic chemotherapy, radiotherapy and laser treatment of EMPD, WLE remains the main-stay of therapy for penoscrotal EMPD.^{4,5} The MD Anderson group performed the largest penoscrotal EMPD series in the United States looking at 20 patients and found that WLE carries the best prognosis provided margins are negative.⁵ A Mayo Clinic study demonstrated improved recurrence rates with Mohs surgery in lieu of WLE but the study had a sampling bias against WLE (83) versus Mohs surgery (12).⁷ The MD Anderson group recommended using Mohs surgery to obtain margins followed by excision of the central tumor. The size and location of our patient's lesion precluded traditional Mohs surgery. Mohs surgery requires specialized training and equipment which are not at the disposal of most urologists. Substituting punch biopsies for the microscopic shave biopsies, however, may offer similar outcomes. The punch biopsies are obtained in a clock-face orientation around the visual border

of the lesion. This facilitates a WLE with negative margins prior to closing what is often a complicated wound in the groin. Our approach to close our patient's complicated wound involved utilizing a vertical rectus abdominis myocutaneous flap (VRAM). VRAM flaps are well vascularized to fill complex perineal defects, provide good cosmetic results, and have been shown to have fewer complications of cellulitis, dehiscence, and infection when compared to thigh flaps.⁸

While many options exist for treatment of penoscrotal EMPD, given that frozen section margins can be unreliable, it is crucial to confirm the final permanent margin status before performing a complicated wound closure. We demonstrate that when traditional Mohs surgery is not feasible, a staged, 'clock-face' modified Mohs approach may yield superior oncologic outcomes when compared to standard WLE. Furthermore, the technique is easily mastered without Mohs training. To be successful with this technique there needs to be a systematic approach to the biopsy schematic with detailed labeling of specimens and seamless communication with the pathologist. Finally, this case report reinforces that clinicians should suspect malignancy when a cutaneous lesion does not respond to commonly used topical agents. □

References

1. Crocker HR. Paget's disease affecting the scrotum and the penis. *Trans Pathol Soc Lond* 1889;40:187-191.
2. Siesling S, Elferink MA, van Dijck JA, Pierie JP, Blokk WA. Epidemiology and treatment of extramammary Paget's disease in the Netherlands. *Eur J Surg Oncol* 2007;33(8):951-955.
3. Moretto P, Nair VJ, Hallani SE et al. Management of penoscrotal extramammary Paget disease: case series and review of the literature. *Curr Oncol* 2013;20(4):e311-e320.
4. Isrow D, Oregel KZ, Cortes J et al. Advanced extramammary Paget's disease of the groin, penis, and scrotum. *Clin Med Insights Oncol* 2014;8:87-90.
5. Hegarty PK, Suh J, Fisher MB et al. Penoscrotal extramammary Paget's disease: the University of Texas M. D. Anderson Cancer Center contemporary experience. *J Urol* 2011; 186(1):97-102.
6. Hashemi P, Kao GF, Konia T, Kauffman LC, Tam CC, Sina B. Multicentric primary extramammary Paget disease: a token cell disorder? *Cutis* 2014;94(1):35-38.
7. O'Connor WJ, Lim KK, Zalla MJ et al. Comparison of Mohs Micrographical surgery and wide excision for extramammary Paget's disease. *Dermatol Surg* 2003;29(7):723-727.
8. Nelson RA, Butler CE. Surgical outcomes of VRAM versus thigh flaps for immediate reconstruction of pelvic and perineal cancer resection defects. *Plast Reconstr Surg* 2009;123(1):175-183.