

# Mohs Micrographic Surgical Approach in a Case of Extramammary Paget's Disease

Kyu Kwang Whang, M.D., Jin Hyoung Won, M.D.

*Department of Dermatology, Ewha Womans University College of Medicine, Seoul, Korea*  
*Department of Dermatology, Yonsei University Wonju College of Medicine, Wonju, Korea*

A 67-year-old man noted a 4×6cm sized, erythematous, erosive, oozing patch on the penile shaft, which resisted conservative therapy for three years. Histopathologic examination revealed typical findings of extramammary Paget's disease. Interestingly, nests of Paget's cells in some hair follicular structures were shown in the middermis. The lesion was excised by microscopically controlled excision, using the fresh-tissue technique. There were histologically involved areas extending to quite a distance beyond the clinically normal appearing skin. Mohs micrographic surgery might offer the unique advantage of effective surgical treatment with microscopically controlled tumor free borders in the extramammary Paget's disease. (*Ann Dermatol* 6:(1) 59~62, 1994)

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**Key Words:** Extramammary Paget's disease, Mohs micrographic surgery

Extramammary Paget's disease was first described on the scrotum and penis by Crocker in 1889.<sup>1</sup> This condition is most commonly seen in the genital, perianal and axillary regions. Histologically, unique large round cells with a large nucleus and broad cytoplasm, so-called Paget cells, scatter or form nests in the epidermis and occasionally invade the dermis. The origin of Paget cells is uncertain. However, some ultrastructural surveys revealed differentiation of Paget cells toward sweat gland apparatus.<sup>2</sup>

Silent peripheral extensions of extensions of extramammary Paget's disease may extend irregularly and unpredictably for long distance beyond the borders of the clinically involved skin.<sup>3</sup> The difficulty of determining an adequate resection margin with the usual surgical procedures results in a high recurrence rate.<sup>4,5</sup> Excision under complete microscopical control might permit the greatest assur-

ance of eradication and preserve the maximal amount of normal tissue.

We performed a Mohs micrographic surgical approach in a case of extramammary Paget's disease, and found the extents histologically to the dermal adnexal structures, far beyond the clinical borders.

## REPORT OF A CASE

A 67-year-old man noted the development of a skin rash on the penile shaft, which had resisted conservative therapy for three years. The lesion was an erythematous, erosive, oozing patch accompanied by an itching sensation on the penile shaft, the scrotum and the pubic area (Fig.1). The area involved measured 4×6cm. The results of the following laboratory tests were within normal limits or negative: complete blood count, urinalysis, liver function test, chest X-ray, electrocardiogram, stool examination and carcinoembryogenic antigen. The result of a sigmoidoscopy examination was normal. Biopsy specimens were taken from the border of the erythematous patch. Routine light microscopy of a specimen from the penile shaft revealed a hyperkeratosis, acanthosis, and nests of Paget's cells which are shown as large

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**Reprint request to:** Kyu Kwang Whang, M.D., Department of Dermatology, Ewha Womans University College of Medicine, 70 Chongro 6ga, Chongroku, Seoul, Korea



Fig. 1. The photograph shows a 4×6cm sized, erythematous, erosive, oozing patch on the penile shaft, the scrotum, and the pubic area, of which the margin is defined with gentian violet.

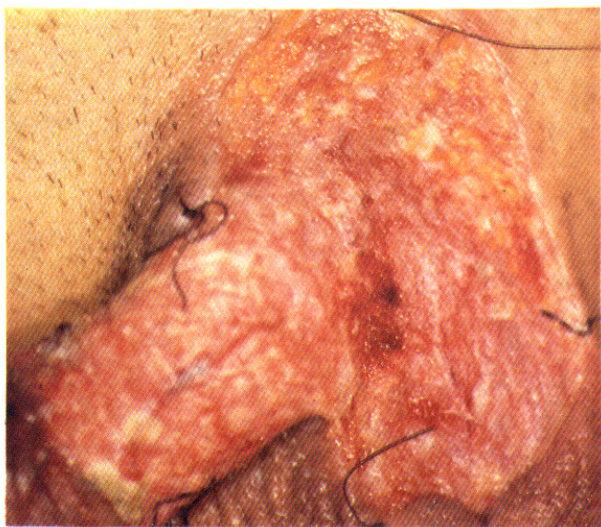


Fig. 3. The lesion was excised 4 to 8mm outside the clinical margin.

pale vacolated cells with hyperchromatic nuclei in the epidermis (Fig. 2). A positive reaction occurred on special staining with diastase-treated PAS. These findings were consistent with a diagnosis of extramammary Paget's disease.

The lesion was excised by microscopically con-

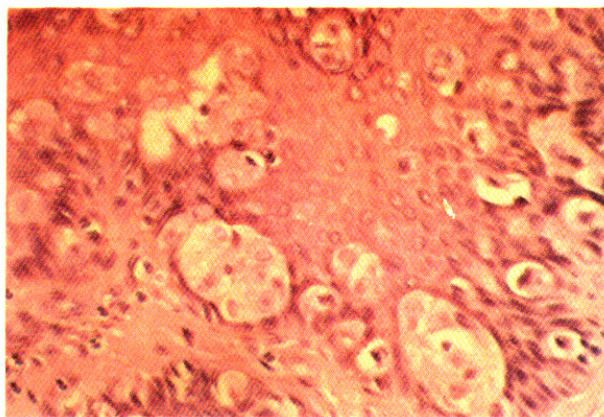


Fig. 2. Routine light microscopy of a specimen from the penile shaft shows a hyperkeratosis, acanthosis, and nests of Paget's cells which were also found outside the clinical margin (H & E, × 200).

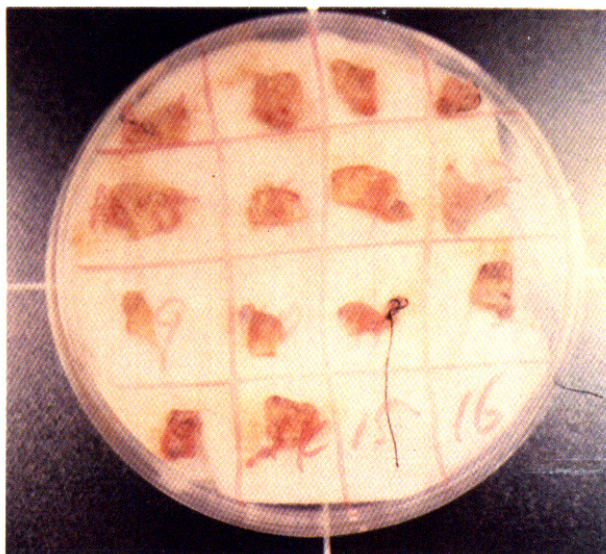


Fig. 4. One half of the involved tissue is divided into 14 pieces.

trolled excision, using the fresh-tissue technique. Before excision, the lesion was divided into two parts (A, B), and each part was excised with No. 15 blade, 4 to 8 mm outside of the clinical margins (Fig. 3). Bleeding control was done with electrocauterization. Each excised tissue was divided from the margin sites and were mapped. One tissue was divided into fourteen pieces and the other tissue was divided into fifteen pieces (Fig. 4). The Paget's cells were found in almost all of the pieces. This finding suggests that the neoplasm extended quite a distance beyond the clinically apparent margins. Nests of Paget's cells existed in the hair



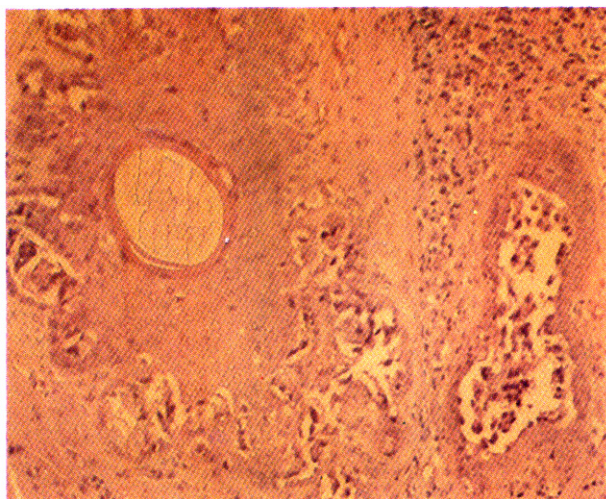


Fig. 5. The photograph shows nests of Paget's cells existed in the outer root sheath cell layer of hair follicle, which was located in the middermis (H & E,  $\times 100$ ).

follicular structures, which were located in the middermis (Fig. 5). These suggest that sufficient depths and widths should be excised whichever treatment modality is chosen. The patient refused the further Mohs surgical stages. The wound healed by second intention (Fig. 6).

## DISCUSSION

Over 50 years ago Frederic E. Mohs developed a technique for the management of cutaneous neoplasms which he called chemosurgery.<sup>6</sup> This technique, which employed serial excisions in conjunction with total histologic control of the margins of resection, yielded high cure rates in patients for whom routine modalities had failed. Mohs micrographic surgery is widely used for excising difficult malignancies of the skin.<sup>7,8</sup> The best indications are persistent tumors, poorly defined margins, aggressive histologic types, high risk anatomic sites or those with high functional or aesthetic value, large neoplasm, or incompletely excised neoplasm.<sup>7</sup>

The essential components of Mohs surgery<sup>7</sup> were 1) tissue excision in a thin layer (often after an initial debulking of the obvious tumor), allowing the lateral and deep margins to be examined in the same plane under the microscope; 2) color coding of tissue with dyes for orientation; 3) tissue mapping using a diagram; 4) horizontally cut frozen sections; 5) examining these sections microscopically to pinpoint the exact locations of

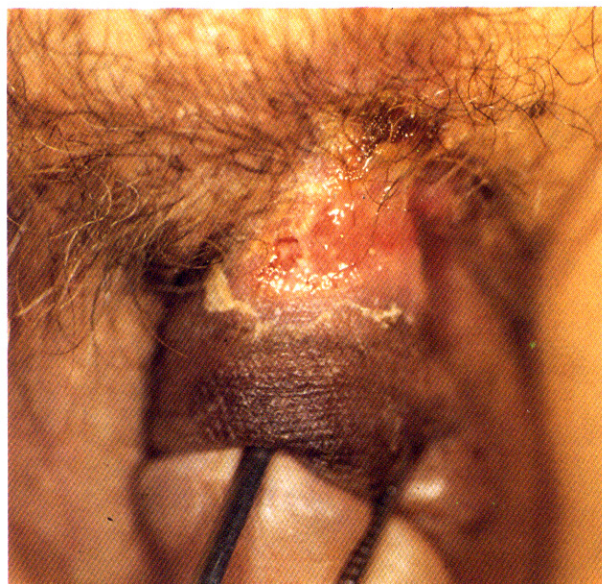


Fig. 6. The wound healed by second intention.

any residual tumors; 6) allowing the wound to heal by secondary intention.

The treatment of extramammary Paget's disease in the past has been primarily surgical.<sup>9-12</sup> Radiotherapy has been tried without success.<sup>13</sup> In the previous review studies, extramammary Paget's disease has a local recurrence rate of 31% to 61%.<sup>4,5</sup> Mohs reviewed 73 cases of extramammary Paget's disease and reported a local recurrence rate of 44%.<sup>3</sup> In 1979, Mohs described his results with the fixed tissue technique in the treatment of five cases of extramammary Paget's disease.<sup>3</sup> All five cases were free of recurrence for periods ranging from four months to nine years. Gunn and Gallagher<sup>14</sup> reported through a topographic study of vulvar Paget's disease that the extent of histologically demonstrable disease was far greater than that of the visible lesion and the outline of the involved area was highly irregular.

Histologic findings showed typical intraepidermal Paget's cells in our case, but quite a few Paget's cells were observed in the outer root sheath cell layers of the hair follicle, which is located in the middermis. Also sections of normal appearing skin 4 to 8 mm from the clinically visible lesions were found to still contain a lot of large Paget's cell nests. The local tumor recurrence may increase due to the difficulty of adequately assessing tumor margins by clinical examination alone. Although our patient had refused the further

Mohs surgical stages, it might have promised the unique advantages of effective surgical treatment with microscopically controlled tumor free borders, while maintaining cosmetic and functional demands by maximal preservation of normal tissue in these conditions. So, we hope to emphasize the advantages of this surgical approach as the preferred treatment modality for extramammary Paget's disease.

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