

Photodynamic Therapy for Primary Cutaneous Extramammary Paget Disease

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Case Presentation

A 70-year-old man presented with a slowly growing erythematous scaly plaque on the right inguinal fold (Figure 1A). The patient complained about itching and burning. Dermatoscopic features observed included a milky-red background with variable vascular patterns, in particular dotted vessels and glomerular vessels with some erosions (Figure 1C). Biopsy showed nests of proliferating atypical cells with pagetoid spread to the entire epidermis (Figure 1E). Cerium ammonium molybdate (CAM) 5.2 stain, marking cytokeratins 8 and to a lesser extent cytokeratin 7, highlighted Paget cells proliferating in the epidermis and upper dermis (Figure 1F). A diagnosis of Extramammary Paget disease (EMPD) was thus made. Thoracic, abdominal, and pelvic computed tomography scan resulted negative. Also, carcinoembryonic antigen (CEA) and prostate-specific antigen (PSA) were in the normal range. The patient received 5 sessions of methyl aminolevulinic acid photodynamic therapy (MAL-PDT) every

2 weeks with a complete resolution of the cutaneous manifestation and without any adverse effects or functional impairments (Figure 1, B and D).

Teaching Point

EMPD is a rare neoplastic disease affecting predominantly patients between 50 to 80 years of age [1]. The current treatments may include surgery, considered the first-line therapy and radiotherapy, imiquimod cream, photodynamic therapy (PDT), topical fluorouracil and carbon dioxide laser therapy. Data for treatment with fluorouracil, carbon dioxide laser, and combination topical therapies are limited. In patients unwilling to undergo surgical treatment or who are not surgical candidates, PDT may represent an interesting approach to EMPD. In particular, MAL-PDT showed complete clearance rate of 27.4 % [2].

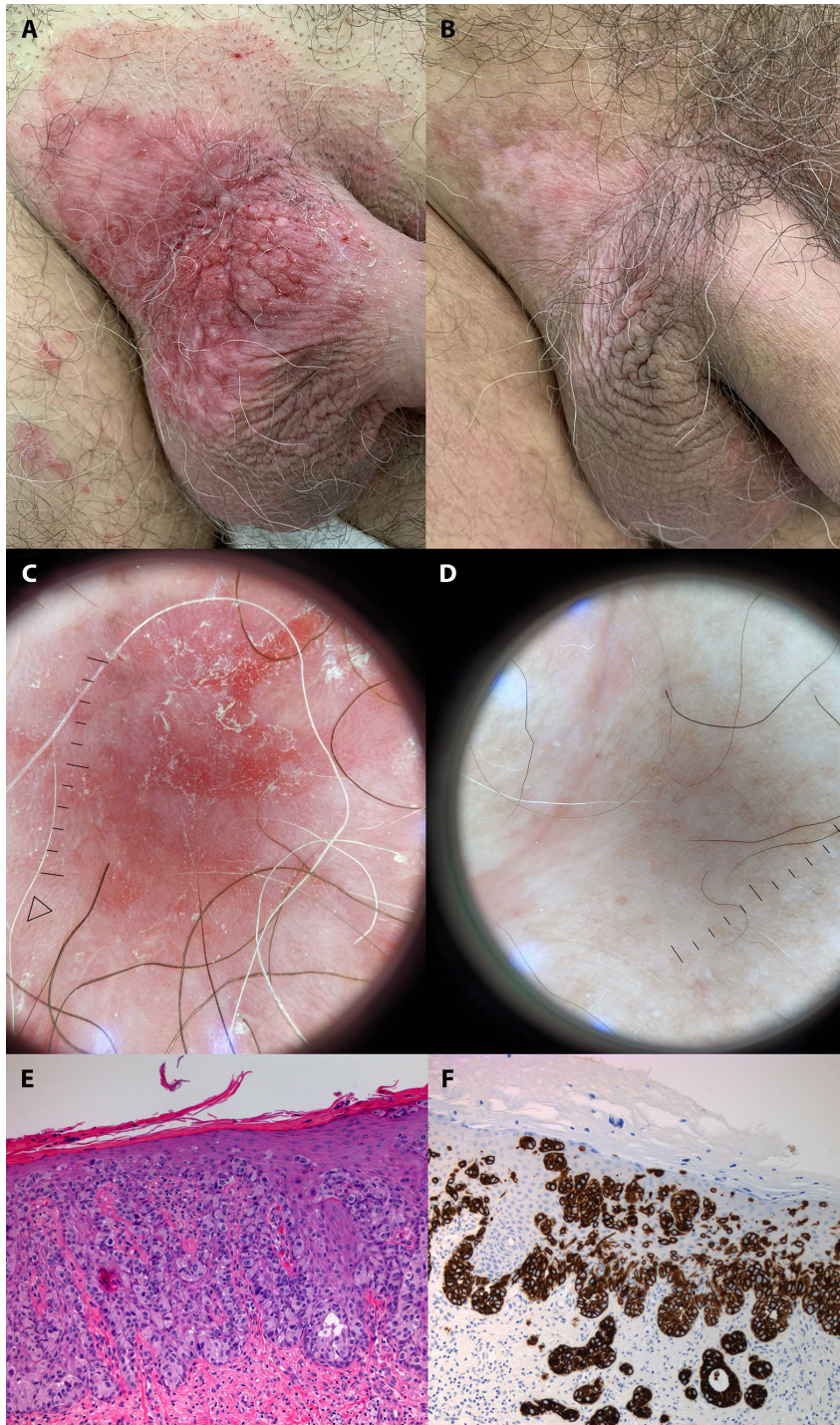


Figure 1. (A) A slowly growing erythematous scaly plaque on the right inguinal fold: a case of EMPD. (B) Resolution of the cutaneous manifestation after five sessions of MAL-PDT. (C) Dermatoscopic features: milky-red background with variable vascular patterns, in particular dotted vessels and glomerular vessels. (D) Dermatoscopic aspects of the skin after MAL-PDT. (E) Hematoxylin and Eosin stained tissue section showed an intraepithelial proliferation of large, round, atypical cells with an abundant, pale mucin-rich cytoplasm (Paget cells), located in nests along the basement membrane and extending to the entire epidermis, with occasional mitoses. Hyperkeratosis and parakeratosis overlie the proliferation, and a dermal infiltrate is evident (H&E, 40×). (F) CAM 5.2 stain marks Paget cells proliferating in the epidermis and upper dermis.

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