

Letter to the Editor

Triple Extramammary Paget Disease in a White Man

To the Editor:

Extramammary Paget disease (EMPD) is an uncommon malignancy that typically presents with 1 or more erythematous patches or plaques in the genital, perianal, or axillary regions.¹ There are rare cases of multisite disease (multiple EMPD) that have occurred mainly in the Japanese population.¹⁻⁵ We report a case of triple EMPD in a white man. A case of triple EMPD in a white patient was reported in the French-language literature.⁶

Case Report

A 76-year-old white man presented to an outside clinic with a pruritic erythematous plaque in the left groin of 1 year's duration. A biopsy was performed and was consistent with EMPD. A comprehensive evaluation by an oncologist, including computed tomography of the chest, abdomen, and pelvis; magnetic resonance imaging of the abdomen and pelvis; colonoscopy; cystoscopy; and laboratory testing, including prostate-specific antigen, revealed no associated internal malignancy.

Soon after, additional biopsies of lesions in the right groin and left axilla were performed, also revealing EMPD. The patient was then referred to our clinic for definitive management.

Our initial examination was remarkable for eczematous plaques on the scrotum and bilateral inguinal folds (Figure 1). There also was a light brownish red patch and smaller light brown macules in the left axilla (Figure 2A) as well as faint brown macules in the right axilla (Figure 2B). Biopsies of lesions in the 3 sites



Figure 1. Eczematous plaques in the right inguinal fold.



Figure 2. Brown macules and patch with a biopsy wound in the left axilla (A) and brown macules in the right axilla (B).

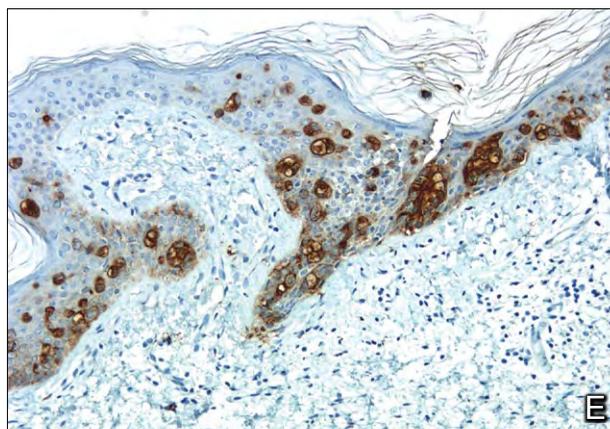
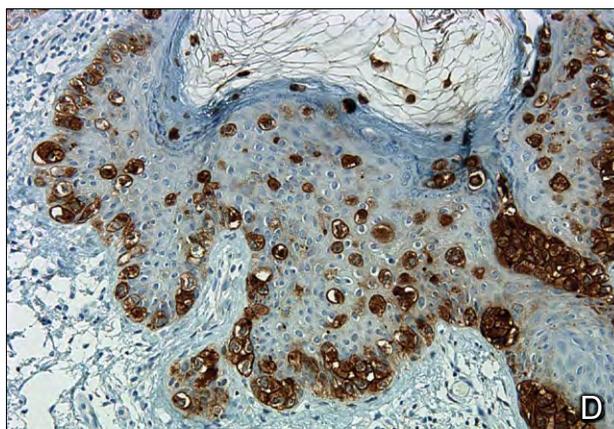
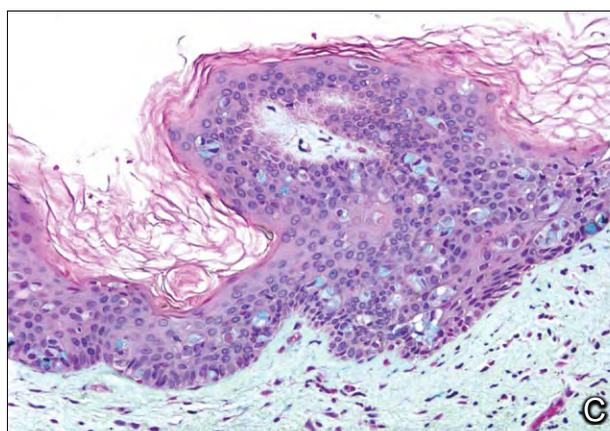
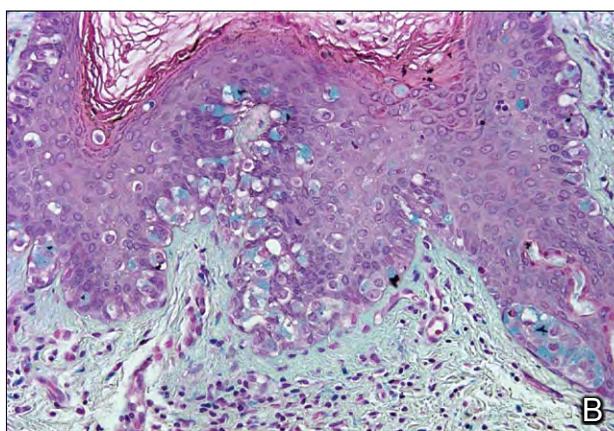
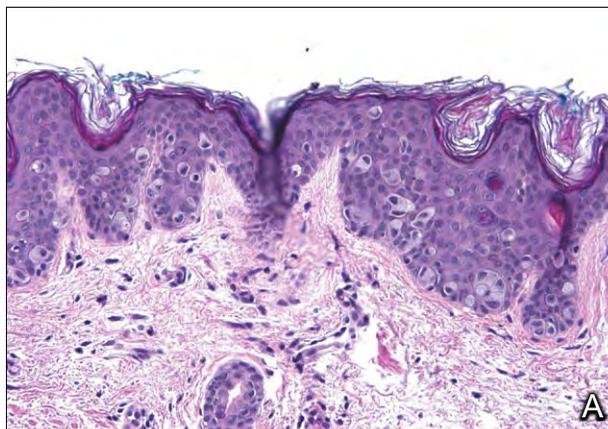


Figure 3. Paget cells distributed singly and in clusters within the epidermis of the right axilla (H&E, original magnification $\times 200$)(A). Paget cells also stained positive with Alcian blue at pH 2.5 in the left axilla (original magnification $\times 200$)(B) and the right axilla (original magnification $\times 200$)(C). Paget cells stained positive with carcinoembryonic antigen in the left axilla (original magnification $\times 200$)(D) and the right axilla (original magnification $\times 200$)(E).

showed similar findings. They all demonstrated numerous Paget cells with abundant pale-staining cytoplasm and large, hyperchromatic, pleomorphic nuclei that were distributed singly and in clusters within the epidermis (Figure 3A). There was no dermal involvement. Tumor cells stained positive with mucicarmine, colloidal iron, Alcian blue at pH 2.5 (Figures 3B and 3C), carcinoembryonic antigen (Figures 3D and 3E),

cytokeratin 7, and cytokeratin AE1/AE3. They were negative for cytokeratin 20, S-100, and Melan-A. The biopsy results were consistent with triple EMPD of primary cutaneous origin of the groin and bilateral axillae.

The patient preferred to have all lesions excised by a plastic surgeon under general anesthesia. Multiple excisions were required to achieve tumor clearance.

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