Multiple-site primary extramammary Paget disease in a 77-year-old man

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A 77-year-old man presented to our dermatology clinic with several erythematous, purpuric, scaling plaques over his genital and suprapubic regions, which had appeared over the previous year (Figure 1A). Similar lesions had developed in both axilla (Figure 1B) over the previous 6 months. He had no history of cancer. Initially, the lesions were assumed to be eczema and then fungal infections, treated with topical steroids and antifungal agents, respectively, without improvement. We conducted punch biopsies from these sites, which showed epithelioid tumour nests and individual cells with epidermal pagetoid spreading (Appendix 1, available at www.cmaj.ca/lookup/doi/10.1503/cmaj.240188/tab-related-content), indicating extramammary Paget disease. Laboratory results were unremarkable. Further examinations — including esophagogastroduodenoscopy, colonoscopy, transurethral ultrasonography, and a computed tomography scan — revealed no internal malignancies, supporting the diagnosis of primary extramammary Paget disease at multiple sites. A plastic surgeon performed wide local excisions with a 2-cm margin for all tumours. We did not observe any recurrence over a 4-year follow-up period.

Mammary Paget disease is characterized by an intraepidermal adenocarcinoma around the areola, often linked to a breast carcinoma. Conversely, extramammary Paget disease is a distinct clinical condition that affects extramammary areas, primarily apocrine gland–bearing skin. Extramammary Paget disease has a higher prevalence among older adults and typically manifests as well-defined, moist, erythematous, and scaly plaques with pigment changes. Frequently affected areas include the vulva (44.8%) and penis or scrotum (27.0%); pruritus is the most common symptom.[1] Although extramammary Paget disease usually manifests at a single site, multiple sites have been documented.[2] Between 5% and 42% of patients with extramammary Paget disease exhibit concurrent extragenital skin cancer or internal malignancies. Differential diagnosis includes chronic eczema, dermatophyte infection, inverse psoriasis, Bowen disease, and cutaneous metastasis from internal malignancies. Clinical identification of extramammary Paget disease poses challenges given its resemblance to other skin lesions. Diagnosis may be delayed by an average of 20 months.[3]

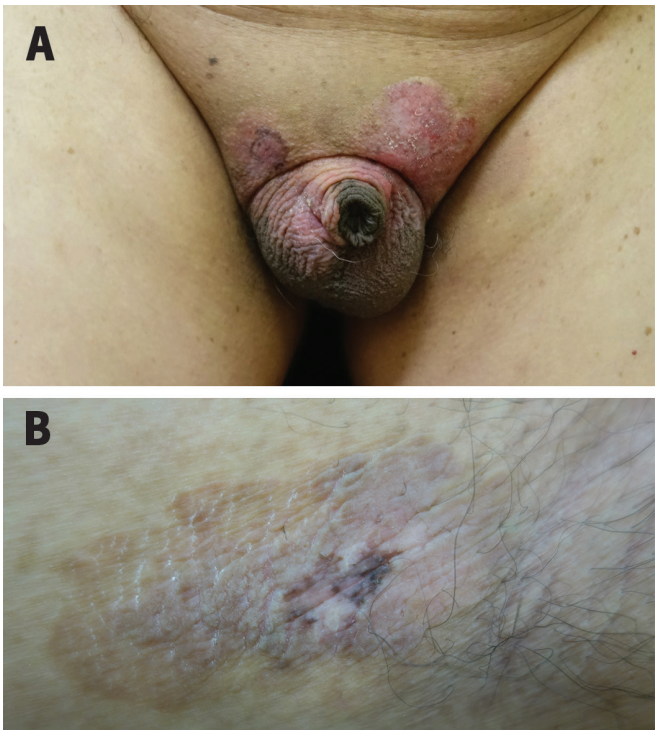

Figure 1: (A) Well-demarcated, erythematous, scaling plaques with central hypo- or hyperpigmentations and shallow erosions over the scrotum, penile shaft, and suprapubic region, and (B) well-demarcated, brownish, scaling plaques with central hypo- or hyperpigmentations over the left axilla in a 77-year-old man with extramammary Paget disease.

References

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