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Extramammary Paget Disease Treated With 5% Imiquimod Cream[☆]

Enfermedad de Paget extramamaria tratada con imiquimod 5% crema

To the Editor:

Extramammary Paget disease (EMPD) is a rare cutaneous neoplasm. It is an intraepithelial adenocarcinoma that develops in areas rich in apocrine glands.¹ The most commonly affected site is the vulva, followed by, in order of frequency, the scrotum, the perianal region, and the axillas.² It is essential to screen for other tumors as EMPD is associated with malignancy at other sites.

A higher prevalence of the disease has been observed in white women aged between 50 and 80 years; because its clinical presentation is variable and nonspecific, EMPD is often confused with dermatitis, leading to delayed diagnosis and treatment.¹

The traditional treatment of choice is surgical excision with wide margins, but even so, the disease has a high rate of recurrence (up to 43%).¹

We describe the case of a 72-year-old woman, with no past history of interest, who consulted for a skin lesion with an eczematous appearance that had appeared on the vulva over a year earlier. Physical examination revealed a scaly erythematous plaque measuring 8 x 5 cm containing whitish islands alongside eroded, exudative areas (Fig. 1). The patient said that she had been treated, without success, with topical corticosteroids, antibiotics, and antifungals for months.

We performed a biopsy of the lesion to confirm the suspected diagnosis of EMPD. Histology showed hyperkeratosis, parakeratosis, acanthosis, and diffuse infiltration by cells with abundant pale cytoplasm and vacuolated nuclei (Paget cells) throughout the epidermis. There was no evidence of dermal infiltration. Immunohistochemistry was positive for carcinoembryonic antigen, epithelial membrane antigen, cytokeratin (CK) 7, and CK18 (Fig. 2).

On confirming the diagnosis of EMPD, we searched for associated malignancy, but the results were negative.

Because of the considerable functional and cosmetic sequelae associated with the surgical treatment of EMPD, we analyzed alternative treatments and opted for 5% imiquimod cream. The patient consented to treatment after having been informed of the good results achieved in cases similar to hers, as well as of the disadvantages associated with this therapy. After cleaning of the area, the cream was applied to the lesion and to 1 to 2 cm of healthy skin around the lesion. Treatment was administered nightly 6 days a week. The patient tolerated the treatment well, despite the initial appearance of erosions and small crusts on the affected area. These lesions did not require interruption of the treatment. The patient was advised to apply fusidic acid ointment to the lesion to prevent superinfection and to help loosen the small crusts. Clinical resolution of the lesion was observed after 6 weeks of treatment (Fig. 3), but the patient was advised to continue treatment up to week 16. After this, the frequency of treatment was reduced to 3 times a week, with monthly outpatient follow-up in the dermatology department up to week 24. A skin biopsy performed several months later confirmed resolution of the disease and confirmed the absence of tumor cells. Once-weekly topical imiquimod was prescribed as maintenance therapy.³

At the time of writing, after 14 months of follow-up, there were no signs of recurrence.



Figure 1 Scaly erythematous plaque containing whitish islands alongside eroded, exudative areas on the vulva.

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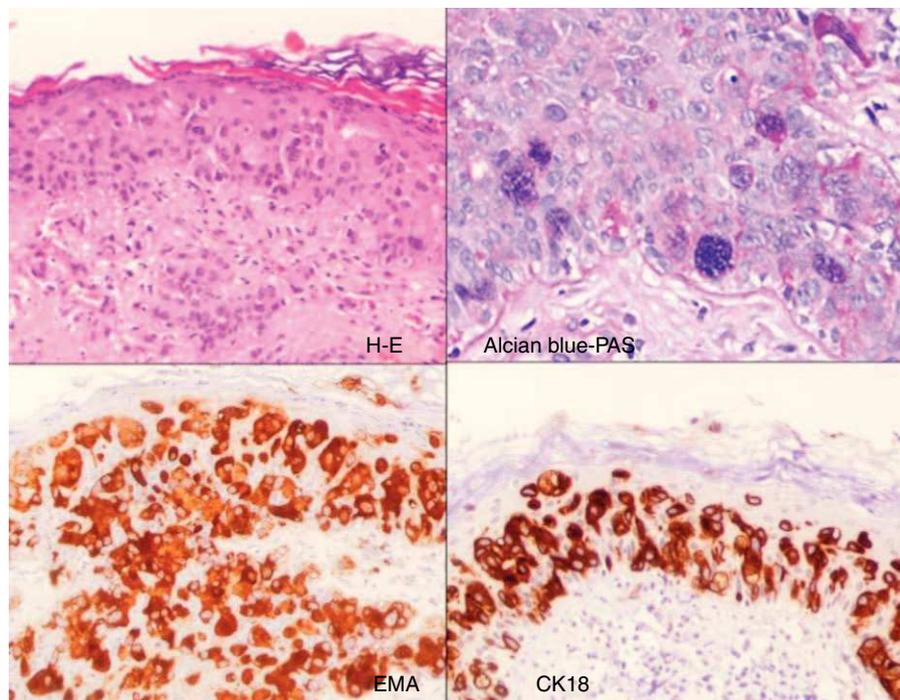


Figure 2 Immunohistochemistry was positive for carcinoembryonic antigen, epithelial membrane antigen (EMA), cytokeratin (CK) 7, and CK18 (original magnification x40). H-E, hematoxylin-eosin; PAS, periodic acid Schiff.

The treatment of choice for EMPD has traditionally been surgical excision with control of margins or Mohs micrographic surgery. In recent years, however, there have been reports of small series and isolated cases in which EMPD has been treated with other methods such as electrodesiccation and curettage, laser therapy, photodynamic therapy with aminolevulinic acid, topical chemotherapy with bleomycin or 5-fluorouracil, and topical 5% imiquimod, all of which have shown the potential offered by nonsurgical approaches.⁴⁻⁷ It should be noted, however, that none of these treatments can be used to assess the persistence of disease. A standardized treatment regimen has not been established for imiquimod. Indeed, the literature contains varying reports of both treatment frequency (from once

daily to 3 times a week) and duration (from 6 to 36 weeks). The maintenance regimen administered in our patient was based on previous recommendations to reduce the high risk of recurrence of the lesion.³

In our opinion, 5% topical imiquimod should be considered in patients with EMPD without dermal infiltration or associated underlying malignancy, and is particularly interesting for large lesions or patients with a high surgical risk.⁴⁻⁸ Its advantages over surgery are its noninvasive nature and its ability to target subclinical lesions. Nonetheless, further controlled studies are needed to confirm the effectiveness and safety of this treatment and to determine the most suitable treatment frequency and duration.



Figure 3 Clinically resolved lesion.

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Acquired Nonfacial Dermal Melanocytosis[☆]

Melanocitosis dérmica adquirida extrafacial

To the Editor:

Dermal melanocytosis includes a wide variety of congenital and acquired, histologically indistinguishable entities characterized by an intradermal proliferation of fusiform pigment-bearing melanocytes in the absence of melanophages.^{1,2} Clinically the lesions appear as brown or bluish macules, depending on whether the melanin pigment is located predominantly in the superficial or deep dermis. Congenital dermal melanocytosis includes Mongolian spots and nevi of Ota and of Ito. Nevus of Ota (nevus fusoceruleus ophthalmomaxillaris) typically develops on the face in the area of distribution of the trigeminal nerve, whereas nevus of Ito (nevus fusoceruleus acromioclavicularis) affects the shoulder and neck region. These nevi, which are present at birth in around 60% of cases, rarely disappear in later years. Mongolian spots occur on the lower back or buttocks and are also usually present at birth, but generally regress in the first years of life.³

Acquired dermal melanocytoses are rare. Hori et al⁴ first described acquired bilateral Ota-like nevus in 1984. Although acquired melanocytoses tend to affect the face, they have also been reported on nonfacial sites (the upper and lower extremities, back, hands, and feet). Prevalence is highest among Asian women, and appearance in white individuals is very rare.⁵

We describe the case of a 49-year-old woman with an 8-year history of an asymptomatic bluish-gray skin lesion, 25 cm in diameter, located on the right side of the upper back (Fig. 1). The patient stated that she was taking no drugs and that she had not experienced any trauma or inflammatory disorders in the affected area. Two years before the consultation, she had undergone surgery for Astler-Coller stage B2 colorectal cancer. Two biopsies of the skin lesion were fixed in formalin and embedded in paraffin for conventional histology study. Both samples revealed a proliferation of pigmented fusiform dermal melanocytes between collagen bands (Fig. 2A). Staining was positive for Melan-A, HMB-45

and S-100 (Fig. 2B). There was no evidence of an increase in epidermal melanocytes or of melanophages in the dermis. These histologic findings confirmed our clinical diagnosis of acquired dermal melanocytosis.

Nonfacial acquired dermal melanocytoses are very rare, particularly in white persons. There have been fewer than 30 cases reported, of which only 3 were in white individuals.⁶ The disorder predominantly affects middle-aged adults, typically Asian females. Malignant transformation of these lesions has occasionally been described.⁷

The pathogenesis of acquired dermal melanocytosis is uncertain. The existence of latent dermal melanocytes resulting from abnormal migration from the neural crest or from the hair bulbs or epidermis (a process known as "dropping off") has been suggested as a possible cause.^{4,8,9} The dormant melanocytes may be reactivated by exogenous agents such as solar radiation, local inflammation, trauma, drugs, hormone therapy with estrogen or progesterone, or other, as yet undefined, stimuli. We could associate none of



Figure 1 Bluish-gray macule on the right side of the back.

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