Case for diagnosis*

Caso para diagnóstico

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CASE REPORT

An 83 year-old man with a history of total bilateral amaurosis, of unknown cause, and with no other relevant known diseases, was observed for an 8 year-long pruritic polycyclic erythematous plaque of the left axilla (Figure 1). He had been continuously medicated with topical antifungals and steroids, with disease progression. No lymphadenopathies were found. Histology of skin biopsy revealed an intraepidermal proliferation of large cells with clear cytoplasm and pleomorphic nuclei, either loosely scattered or in small nests (Figure 2). Immunohistochemistry was CK7 positive and negative for CK20 and Melan A (Figure 3). After diagnosis of Extramammary Paget's Disease, total excision with 2cm margins was performed, with a posterior brachial cutaneo-fascial flap and a graft from the right thigh. Mammography, breast ultrasound and thoraco-abdomino-pelvic computerized tomography detected no changes. 24 months after surgery the patient remains without evidence of local or underlying internal malignancy.

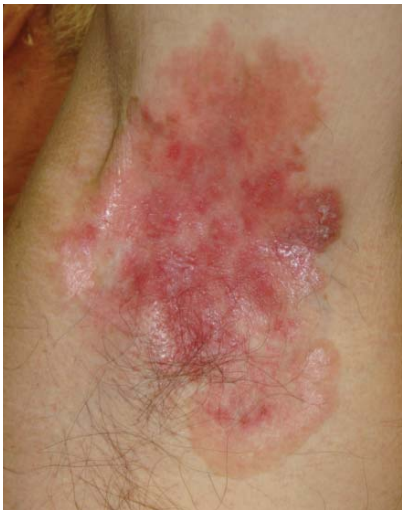

FIGURE 1: Isolated erythematous plaque with focal desquamation, erosions, and grossly polycyclic features

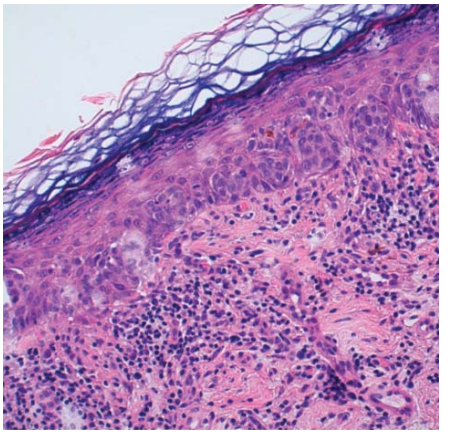

FIGURE 2: Intraepidermal infiltrate of large cells with clear cytoplasm and pleomorphic nuclei, either loosely scattered or in small nests (H&E × 200)

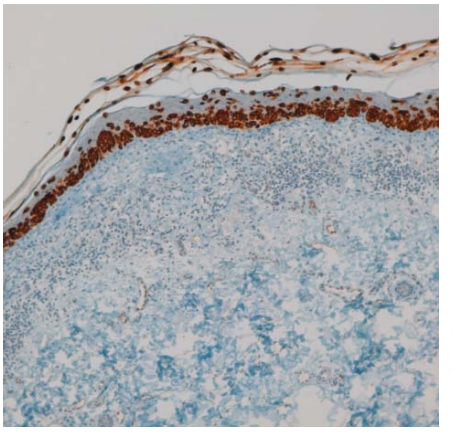

FIGURE 3: Immunohistochemical staining for cytokeratin 7 demonstrating notorious pagetoid spread (CK7 × 100)

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DISCUSSION

EMPD is a rare malignant intraepidermal neoplasm, morphologically and histologically similar to Mammary Paget's Disease, where the main difference is localization.[1] Its histiogenic origin remains under study, although most cases represent a primary intraepidermal adenocarcinoma (primary EMPD). A small number of cases probably arise from intraepithelial extension of cutaneous adnexal cancers or from a regional internal neoplasm with contiguous epithelium (secondary EMPD).[2]

Clinically, lesions are unspecific and mistakenly diagnosed and treated for several years before a diagnosis of EMPD. It manifests typically as a well-defined, solitary, scaly and erythematous plaque with eventual erosions, exudation or nodules, characteristically affecting apocrine gland rich areas (the groin or, more rarely, the axilla).[2,3] The differential diagnosis includes: eczema, psoriasis, superficial mycosis, basal cell carcinoma, Bowen's disease, and malignant melanoma (especially when pigmented).

Histologically, the diagnosis of pagetoid spread typically observed includes Melanoma, Bowen's Disease, Langerhan Cell Histiocytosis, Mycosis Fungoides, Sebaceous Carcinoma and Merkel Cell Carcinoma.[2]

An association with cancers of the breast, bladder, prostate, vulva and gastrointestinal tract has been reported.[2] The exact incidence is unknown however, due to difficulties that include confirming results between different studies, and making a distinction between those associated with EMPD and EMPD-independent age-expected cancers. A guided investigation may however be programmed, according to the most frequent specific cancer association with the topographical location of EMPD.

In non-invasive EMPD, surgery is still the therapeutic method of choice. Relapses are frequent, although Mohs micrographic surgery guarantees lower relapse rates than conventional wide-margin surgery.[3,4] Photodynamic therapy has been used in EMPD patients. This has advantages from a practical, conservative point-of-view, with a possibility of retreatment, although relapse rates are comparable to those of conventional surgery.[5] We can also point to radiotherapy, CO_2 or Nd:YAG lasertherapy, and topicals such as 5-fluorouracil, imiquimod and bleomycin which may play a part in either the neoadjuvant or adjuvant treatment of the primary cancer or relapses.[6,7,8,9] ❑

Abstract: An 83 year-old man was observed for an 8 year-long pruritic, erythematous plaque of the left axilla. He had been continuously medicated with topical antifungals and steroids with occasional symptomatic relief, but with disease progression. After a clinical and histological diagnosis of Extramammary Paget's Disease, a total excision with wide margins was performed. He remains disease-free 12 months after surgery. We report this rare presentation of Extramammary Paget's Disease on a male patient.
Keywords: Aged; Axilla; Male; Paget disease, extramammary

Resumo: Um homem de 83 anos foi observado por placa eritematosa pruriginosa da axila esquerda com 8 anos de evolução. Estivera continuamente medicado com antifúngicos e corticóides tópicos com alívio sintomático ocasional, contudo com progressão da doença. Após diagnóstico clínico e histológico de Doença de Paget Extramamária, foi feita excisão total com margens largas. Mantém-se sem evidência de doença aos 12 meses após cirurgia. Salientamos esta apresentação rara de Doença de Paget Extramamária num paciente do sexo masculino.
Palavras-chave: Axila; Doença de Paget extramamária; Idoso; Masculino

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