

Case for diagnosis^{*}

Caso para diagnóstico

André Lencastre¹Joana Cabete²Maria Margarida Apetato³DOI: <http://dx.doi.org/10.1590/abd1806-4841.20132311>**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

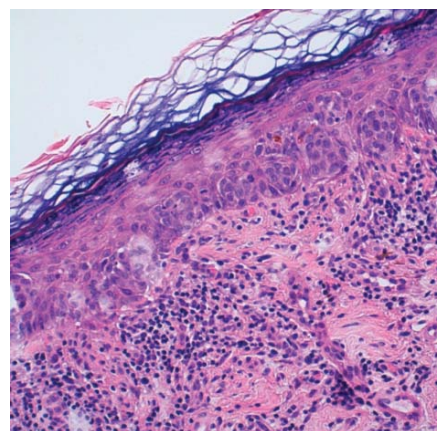


FIGURE 2: Intraepidermal infiltrate of large cells with clear cytoplasm and pleomorphic nuclei, either loosely scattered or in small nests (H&E × 200)

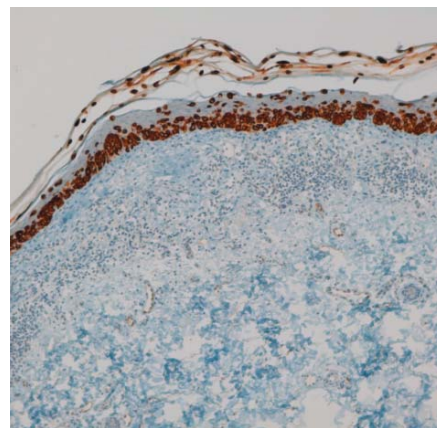


FIGURE 3: Immunohistochemical staining for cytokeratin 7 demonstrating notorious pagetoid spread (CK7 × 100)

Received on 24.11.2012.

Approved by the Advisory Board and accepted for publication on 15.01.2013.

^{*} Work performed in the Dermatology Department, Hospital de Santo António dos Capuchos, Central Lisbon Hospital Center, EPE - Lisbon, Portugal.

Conflict of interest: None

Financial funding: None

¹ Degree in Medicine, Resident Physician in the Dermatology Department, Hospital de Santo António dos Capuchos, Central Lisbon Hospital Center, EPE - Lisbon, Portugal.² Masters Degree in Medicine, Resident Physician in the Dermatology Department, Hospital de Santo António dos Capuchos, Central Lisbon Hospital Center, EPE - Lisbon, Portugal.³ Degree in Medicine, Assistant Physician with Degree in Dermatology, Dermatology Department, Hospital de Santo António dos Capuchos, Central Lisbon Hospital Center, EPE - Lisbon, Portugal.

©2013 by Anais Brasileiros de Dermatologia

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, CO₂ 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

REFERENCES

1. Lage D, Volpini Cde A, Sasseron Mda G, Daldon P, Arruda L. Paget's disease: the importance of the specialist. *An Bras Dermatol*. 2010;85:365-9.
2. Lam C, Funaro D. Extramammary Paget's disease: Summary of current knowledge. *Dermatol Clin*. 2010;28;4:807-26.
3. Chiu C, Yang C, Chen C. Extramammary Paget's disease of the unilateral axilla: a review of seven cases in a 20-year experience. *Int J Dermatol*. 2011;50:157-60.
4. Lee K, Roh M, Chung W, Chung K. Comparison of Mohs micrographic surgery and wide excision for extramammary Paget's Disease: Korean experience. *Dermatol Surg*. 2009;35:34-40.
5. Nardelli A, Stafinski T, Menon D. Effectiveness of photodynamic therapy for mammary and extra-mammary Paget's disease: a state of the science review. *BMC Dermatol*. 2011;11:13.
6. Hata M, Omura M, Koike I, Wada H, Miyagi E, Tayama Y, *et al*. Role of radiotherapy as curative treatment of extramammary Paget's disease. *Int J Radiat Oncol Biol Phys*. 2011;80:47-54.
7. Mengjun B, Zheng-Qiang W, Taseem MM. Extramammary Paget's Disease of the perianal region: a review of the literature emphasizing management. *Dermatol Surg*. 2013;39:69-75.
8. Pang J, Assaad D, Breen D, Fialkov J, Antonyshyn O, Balogh J. Extramammary Paget disease: review of patients seen in a non-melanoma skin cancer clinic. *Curr Oncol*. 2010;17:43-5.
9. Tanaka VD, Sanches JA, Torezan L, Niwa AB, Festa Neto C. Mammary and extramammary Paget's disease: a study of 14 cases and the associated therapeutic difficulties. *Clinics (Sao Paulo)*. 2009;64:599-606.

MAILING ADDRESS:

André Lencastre
Alameda Santo António dos Capuchos
1169-050 - Lisbon
Portugal
E-mail: lencastre.derm@gmail.com

How to cite this article: Lencastre AJG, Cabete J, Apetato M. Case for diagnosis. A lasting axillary plaque on an elderly man. *An Bras Dermatol*. 2013;88(5):828-30.