ECRONICON OPEN ACCESS

EC GYNAECOLOGY
Case Report

Mammary Paget Disease: Case Report and Review of the Literature

Khadija Elboukhari*, Kaoutar Achehboune, Sara Elloudi, Hanane Baybay and Fatima Zahra Mernissi

Department of Dermatology, University Hospital of Fez, Morocco

***Corresponding Author**: Khadija Elboukhari, Department of Dermatology, University Hospital of Fez, Morocco.

Received: January 27, 2020; **Published:** February 25, 2020

Abstract

The mammary Paget disease is an uncommon disorder of the nipple-areola area, it is an intraepithelial adenocarcinoma that is clinically confused with inflammatory or infectious skin disease, this entity is usually underlying a ductal carcinoma. The histology examination identifies Pagets cells infiltrating the epidermis without invasion. We report a case of a 65 years old single woman who presented a Mammary Paget disease with a pemphigus-like appearance on the histology examination. We will also discuss this pathology through a brief review of the literature.

Keywords: *Mammary; Paget; Pemphigus-Like; Ductal Carcinoma*

Abbreviations

MPD: Mammary Paget Disease; ACR: American College of Radiology

Introduction

Mammary Paget disease (MPD) is an uncommon disorder of the nipple-areola area, it is an intraepithelial adenocarcinoma that is clinically confused with inflammatory skin disease [1]. The pathogenesis remain unclear with theories supporting carcinoma de novo or underlying carcinoma [1]. The histology of Paget disease is characterized by neoplastic cells with glandular differentiation [2].

Patient Observation

Patient of 65 years, having as antecedents a pleurisy 40 years ago, which presents since 08months a pruriginous and painless lesion of the right nipple. The dermatological examination found a retracted nipple with an eczematous plaque surmounted by a large crust, that after removing it out we found a very limited ulceration (Figure 1). Dermoscopy examination revealed pink structure less areas, white structureless areas, white scales and a few dotted vessels (Figure 2).

In front of this ulcerated plate was evoked a mammary Paget disease, an epidermoid carcinoma or an achromic melanoma.

Breast ultrasound and mammography concluded that the right nipple was retracted and tumefied with microlobulated and fuzzy posterior contours, with the presence of contralateral breast microcalcifications classified as ACR IV.

A cutaneous biopsy was performed on the ulcerated nipple, showing a basal dystrophic keratinocyte layer mixed with atypical epithelial cells with abundant cytoplasm and large macronucleated hyperchromatic nuclei showing mitotic figures (Figure 3). Without evidence of tumor invasion. Some of these elements float within dehiscences and fissures. The underlying dermis is highly oedematocongestive

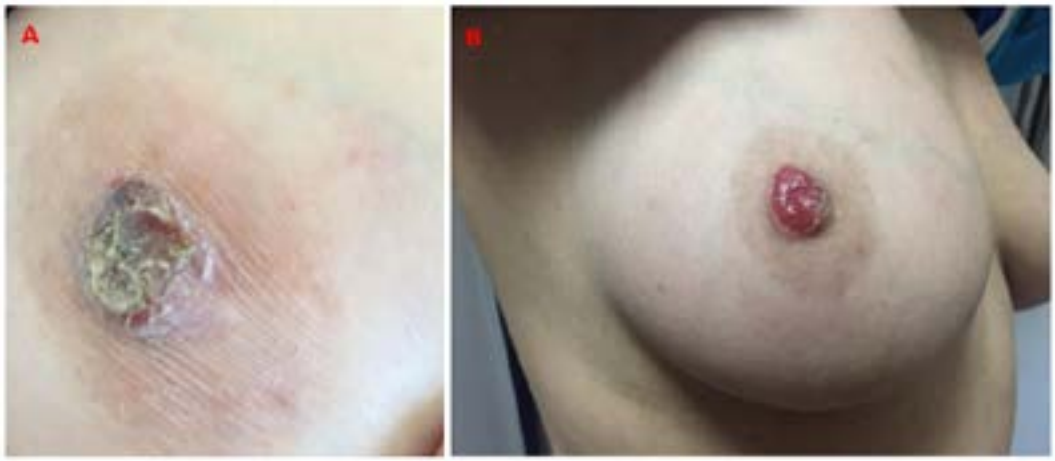

Figure 1: *A: An ecezma like plaque. B: Demarqued ulceration of the retracted nipple.*

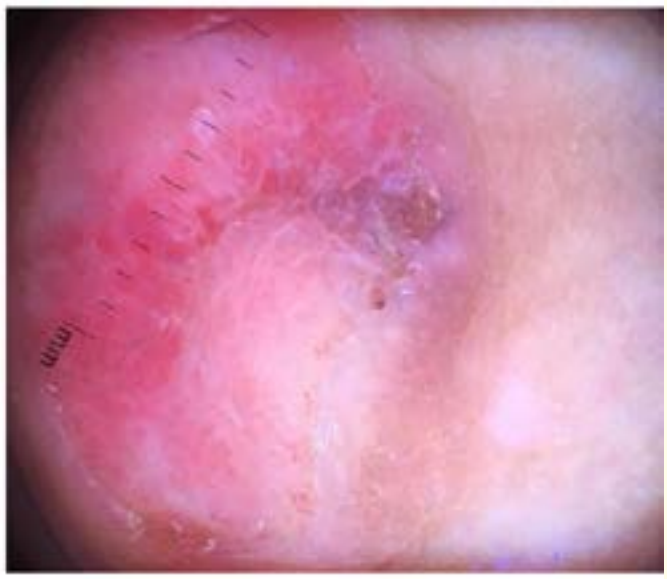

Figure 2: *Pink structure less areas, white structure less areas, white scales and a few dotted vessels.*

seat of an essentially mononucleate dense inflammatory infiltrate. An immunohistochemical complement confirmed the Paget's disease by showing an intense and diffuse expression of the anti-CK7 (Figure 4) antibody and the anti-CK8/18 antibody in addition to a strong membrane labeling. More than 90% of the tumor cells by the anti-c-erbB-2 antibody (Figure 5): score 3+. And finally an heterogeneous expression of the anti-GCDFP15 antibody.

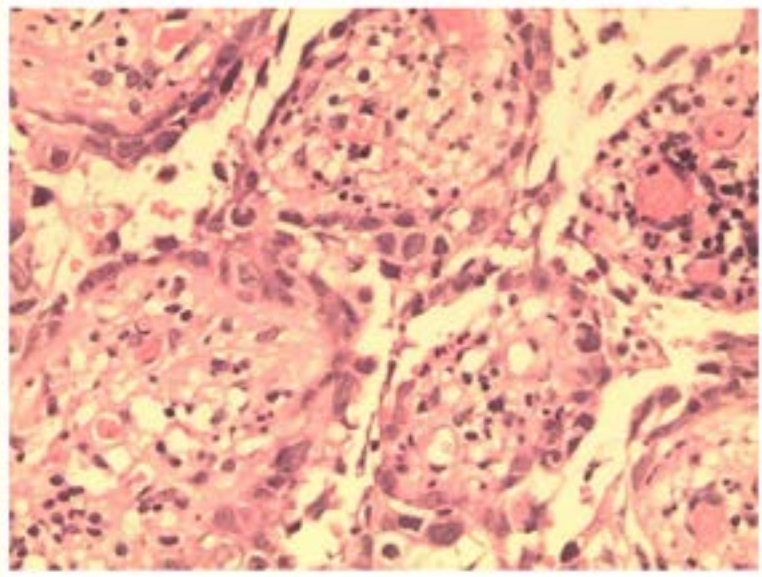

Figure 3: *G x 400 HES coloration-atypical cells in the fissurations.*

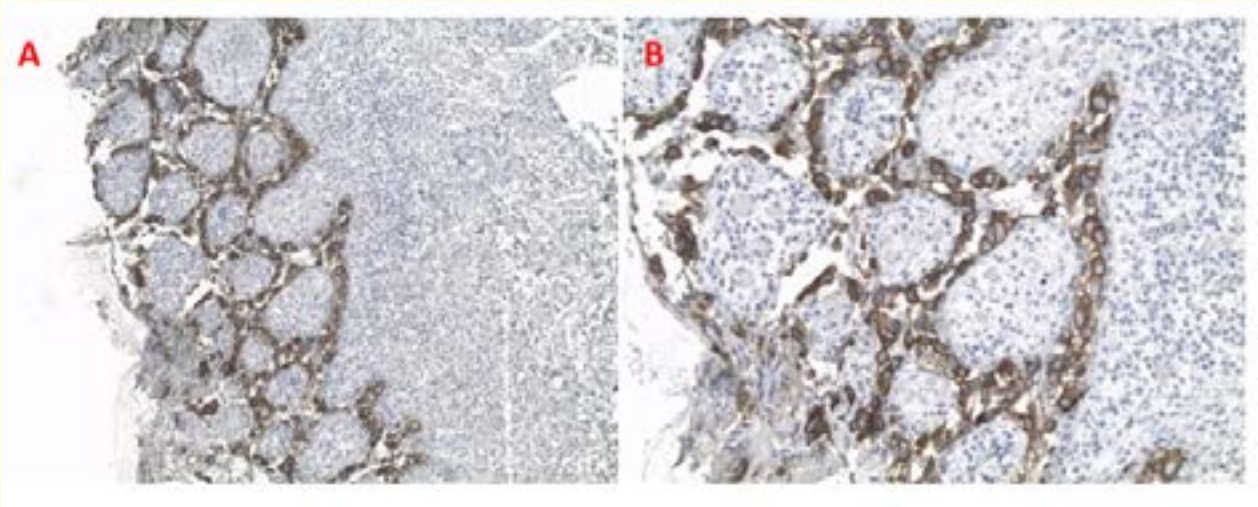

Figure 4: *A: G x 100. Intense and diffuse expression of CK7. B: G x400. Intense and diffuse expression of CK7.*

Our patient had a right mastectomy with histological analysis showed an extensive high-grade carcinoma in situ, with an a cribriform architecture and necrosis. The patient also benefited from a tumerectomy of the left breast after locating the micro-calcifications, whose anatomopathological study did not show any malignancies.

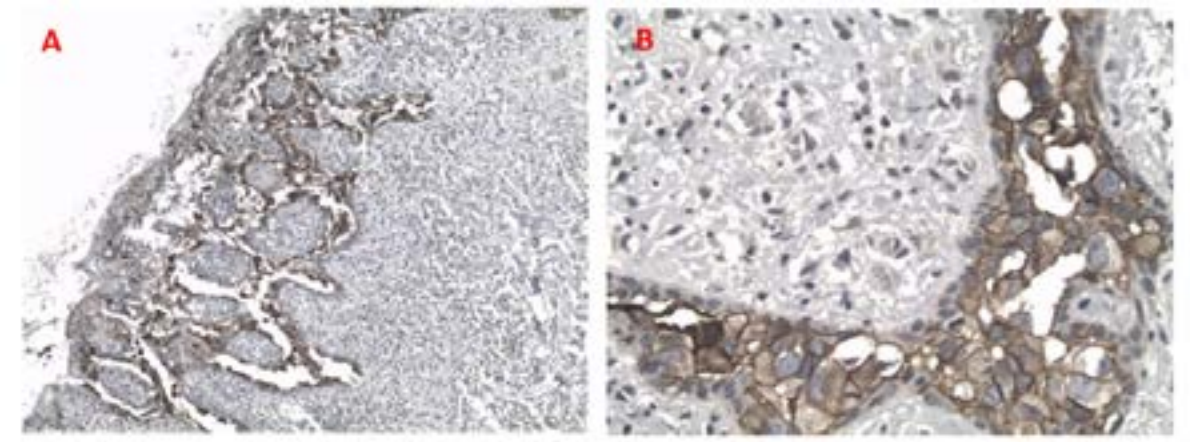

Figure 5: *A: Cerb 2 G x 100 complete membrane labeling with more than 90% of tumor cells. B: Cerb 2 G x 400. Complete membrane labeling with more than 90% of tumor cells.*

Discussion

Presenting 1% of all breast cancers [3], MPD is an intraepithelial adenocarcinoma of the nipple that reveals an invasive adenocarcinoma in 90% of cases [4,5]. This carcinoma in situ was first described in 1874 by Sir James Paget [6,7]. The classic clinical presentation is an eczematous patch or plaque extending from the nipple to the surrounding areola and breast. Functional signs may be pruritus, pain or burning sensation, these signs may precede the clinical appearance of the disease [8,9]. MPD affects an elderly population [1]. Although nipple involvement is pathognomonic of the disease, it may be absent in some cases [10].

The most frequent dermoscopic criteria of MPD are white lines, dotted vessels pink structureless areas, white scales, erosion or ulceration [1]. Those criteria were either found in our patient's lesion. Amongst non-pigmented lesions of the nipple, Contact dermatitis is the most common differential diagnosis of MPD and dermoscopy might enhance the differentiation between these two entities. Contact Dermatitis is dermoscopically characterised by the combination of scattered dots vessels and yellow scales [11] which is absent in MPD [1].

Histologically Paget's disease is characterized by the presence of neoplastic cells scattered throughout all layers of the epidermis [12]. Pagets cells originating from the luminal lactiferous ductal epithelium [13] are typically pale with large nuclei showing a single prominent nucleoli, their cytoplasm is pale and abundant [10]. What was atypical in our observation, is the presence of large fissurations between Paget cells, mimicking a pemphigus (pemphigus-like appearance)

Immunohistochemical complement confirm the diagnosis with a fixation of CK7 and cerb-2 [10,14] MPD management is still a topic of debate; in fact, it proved that a conserving therapy with lymphectomy and radiotherapy is an effective therapy for MPD associated with an in situ or invasive ductal carcinoma of the breast compared to the mastectomy [14].

Conclusion and Perspective

MPD is a rare pathology of the nipple often associated to a ductal carcinoma. The histology is characterized by the presence of atypical cells dissociating the epidermis. The pemphigus like aspect is secondary to this dissociation was clear in our case and may be found in several cases. This will be evaluated by a prospective study and defined the subject of our next work.

Bibliography

1. Sandoval-Leon AC., *et al.* "Paget's disease of the nipple". *Breast Cancer Research and Treatment* 141 (2013): 1-12.
2. Lloyd J. "Mammary and extramammary Paget's disease". *Journal of Clinical Pathology* 53 (2002): 742-749.
3. Apalla Z., *et al*. "Dermoscopic features of mammary Paget's disease: a retrospective case-control study by the International Dermoscopy Society". *The Journal of the European Academy of Dermatology and Venereology* (2019): 15732.
4. Waldman RA., *et al*. "Skin diseases of the breast and nipple: benign and malignant tumors". *Journal of the American Academy of Dermatology* 80 (2019): 1467-1481.
5. Gaurav A., *et al*. "Practical consensus recommendations for Paget's disease in breast cancer". *South Asian Journal of Cancer* 7 (2018): 83-86.
6. Paget J. "On disease of the mammary areola preceding cancer of the mammary gland". *Bartholomew's Hospital Report* 10 (1874): 87.
7. Thin G. "On the connection between disease of the nipple and areola and tumors of the breast". *Transactions of the Pathological Society of London* 32 (1881): 218.
8. Marshall JK., *et al*. "Conservative management of Paget's disease of the breast with radiotherapy: 10- and 15-year results". *Cancer* 97.9 (2003): 2142-2149.
9. Chaudary MA., *et al*. "Paget's disease of the nipple: a ten year review including clinical, pathological and immunohistochemical findings". *Breast Cancer Research and Treatment* 8.2 (1986): 139-146.
10. Broecker JS., *et al*. "An atypical presentation of Paget's Disease of the breast without nipple involvement: Case report and review of the literature". *Pathology - Research and Practice* 213 (2017): 1454-1456.
11. Lallas A., *et al*. "Dermoscopy in general dermatology". *Dermatologic Clinics* 31 (2013): 679-694.
12. Guitera, P., *et al*. "Reflectance confocal microscopy for diagnosis of mammary and extramammary Paget's disease". *The Journal of the European Academy of Dermatology and Venereology* 27 (2013): e24-e29.
13. Yao Y., *et al*. "Breast-Conserving Surgery in Patients With Mammary Paget's Disease". *Journal of Surgical Research* 241 (2019): 178-187.
14. Ooi PS., *et al*. "Mammary Paget's Disease of the Nipple: Relatively Common but Still Unknown to Many". *Journal of the Korean Academy of Family Medicine* (2018).

Volume 9 Issue 3 March 2020