

Paget's Disease of the Nipple a Case Study with Literature Review

Youssef Janati, Abraham Alexis Sanoh*, Edith Ngawa Ngalande, Fatima Zohra Fdili Alaoui, Sofia Jayi, Hikmat Chaara and Moulay Abdelilah Melhouf

University Teaching Hospital Fez, Sidi Mohamed Ben Abdellah University of Fes, Morocco

***Corresponding Author:** Abraham Alexis Sanoh, Résident in Gynecology and Obstetrics, University Teaching Hospital Fez, Sidi Mohamed Ben Abdellah University of Fes, Morocco.

Received: March 10, 2024; **Published:** March 26, 2024

Abstract

The clinical examination of nipple-areolar plaques is an integral part of breast screening evaluation. Any persistent unilateral lesion of the nipple should warrant particular attention and investigation for Paget's disease of the nipple. Its diagnosis is made by cytological scraping of the nipple. This is a rare variant of ductal carcinoma *in situ*. Breast evaluation should look for underlying breast tumors, present in more than 80% of cases, as well as multifocal tumors, which occur with high frequency. Preoperative breast MRI is useful if conservative surgery is being considered due to a higher incidence of occult cancer on mammography and ultrasound. Erosive adenomatosis of the nipple is benign but can mimic pure Paget's disease of the nipple. More rarely, pagetoid basal cell carcinoma, Bowen's disease and melanoma are clinically difficult to differentiate and share with Paget's disease the unilateral nature and chronicity of the evolution. The biopsy allows the diagnosis to be made. Skin extension of breast carcinoma is rare and should be suspected in cases of nipple retraction and/or fixed nipples. Eczema is different in the bilateral nature of its lesions, the absence of deformation of the nipple, the progression by flare-ups and its regressive nature under local corticosteroid therapy.

Keywords: *Paget's Disease of the Nipple; Symptoms; Therapeutic Attitude; Clinical Case*

Introduction

Paget's disease of the nipple is a rare condition that accompanies 1 to 4% of breast cancers. It corresponds to the infiltration of the epidermis of the nipple by adenocarcinoma-type cells. Clinically, it results in eczematous eruptions on the nipple and areola. The association of Paget's disease with ipsilateral breast cancer is found in 82 to 100% of cases. The benefit of imaging based on echo mammography and breast magnetic resonance imaging (MRI) is therefore to search for underlying cancer.

Its management, not yet consensual, varies between radical treatment and removal of the Nipple Areola Complex ("NACectomy"). This is caused by the accompanying breast cancer.

This case report and a review of the literature, this article aims to clarify the clinical particularities and the therapeutic management of this disease.

Case Report

A 40-year-old multiparous patient with no family history of cancer was referred to us from the dermatology department for nipple erosion. This lesion had started two years ago in the form of a single-pore bloody discharge associated with a scaly plaque affecting the nipple and areola of the right breast. Faced with the rapid evolution of the lesion over the last month, the patient was referred to us for treatment.

Clinical outcome

Clinical examination found an erythematous peri-nipple lesion surmounted by a small erosion without discharge on nipple pressure or palpable nodule (Figure 1). The remainder of the left breast examination was unremarkable.

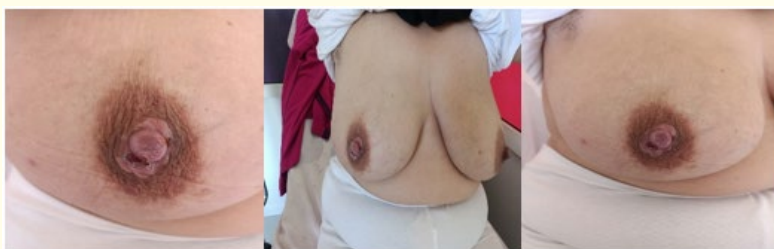


Figure 1

Diagnostic approach

Mammography plus breast ultrasound found a tissue lesion classified ACR4C measuring 3.8 x 2.7 mm in the right breast below the nipple associated with two lymph nodes, one right axillary with thickened cortex and the other intra-mammary lower inner quadrant of the right breast classified ACR2; two retro-nipple cystic lesions of the upper outer quadrant classified ACR2 and ACR3, the anatomo-pathology result found an epidermal proliferation of pagetoid appearance first suggesting Paget's disease confirmed by immunohistochemistry which confirmed the diagnosis of Paget's disease, cytopuncture of the right axillary lymph node returned in favor of a substantially normal lymph node parenchyma.

Breast MRI found a discreet thickening with early enhancement of the nipple and the peri-nipple skin covering in relation to her Paget's disease, thus ruling out the presence of the ACR4c lesion objectified on echo mammography, associated with a right axillary lymph node with thickened cortex including the cytopuncture came back benign. The etiological assessment was unremarkable. The final diagnosis of Paget's disease was made.

Therapeutic intervention and follow-up

The patient benefited from NACectomy type breast surgery, while preserving the mammary gland which made it possible to remove the entire tumor with the definitive anatomo-pathology histological appearance of PAGET disease, proliferation at a distance from the resection limits and the breast parenchyma is the site of foci of florid ductal hyperplasia without having seen carcinoma *in situ* (CIS) or foci of tumor invasion and placed under radiotherapy with good clinical evolution.



Figure 2

Discussion and Conclusion

Paget's disease of the nipple is isolated in 1.4 to 13.3% of cases and associated with ipsilateral breast cancer in 82 to 100% of cases; 13.3 to 52% of these cancers are carcinomas *in situ* and 30 to 60% are invasive carcinomas [3]. Imaging is very useful in diagnosing underlying breast cancer, with mammography having a sensitivity of 97% for detecting tumors in the presence of a palpable mass, but only 50% for detecting tumors in the absence of a palpable mass. MRI is more sensitive than mammography for the *in-situ* diagnosis of an associated carcinoma. An MRI should be performed when mammography and ultrasound are normal [4,5]. Diagnosis Paget's disease is confirmed by histological examination. There is a tumor proliferation of large round cells in the epidermis with rich, pale cytoplasm and a hyperchromatic and nucleolated nucleus. However, this histological appearance is not specific and can be suggestive of other types of skin tumors such as Bowen's disease or melanoma. Paget's disease cells usually express low molecular weight cytokeratins, including CK7, this marking allowing, with a sensitivity close to 100% of confirm the diagnosis. However, in very rare cases, the staining for CK7 can be negative [6]. The main differential diagnosis of Paget's disease is nipple eczema. But the unilaterality, the scalability and the absence of response to corticosteroid therapy make it possible to correct the diagnosis.

Other differential diagnoses may also be considered, such as psoriasis, superficial basal cell carcinoma, and melanoma, which can only be confirmed by histology. For solitary Paget's disease of the breast, without an underlying palpable mass, conservative treatment (resection of the NAC followed by radiotherapy to the entire breast \pm overdose) is necessary [7]. Treatment for cancer-related Paget's disease depends in part on the underlying cancer. Conservative surgery combines a bloc resection of the nipple-areola complex and the underlying tumor, followed by radiotherapy if Paget's disease allows the underlying tumor to be removed by NAC without severe aesthetic sequelae [8]. However, a total mastectomy may still be necessary if Paget's disease associated with multifocal cancer precludes one-piece resection, or if the tumor is large or a contraindication to radiotherapy. Performing sentinel lymph nodes is recommended in cases of underlying invasive cancer. It does not seem necessary in patients with a non-invasive central tumor, especially if the MRI of the remaining lymph nodes is normal [9]. Paget's disease of the breast, alone or associated with adenocarcinoma *in situ*, has a rate of 15-year survival of almost 90% and when associated with invasive adenocarcinoma, a 15-year survival rate of 60% [10].

Bibliography

1. Paget J. "On disease of mammary areola preceding cancer of the mammary gland". *ST. Bartholomew's Hospital Report* 10 (1874): 87-89.
2. Chen CY, *et al.* "Paget's disease of the breast: changing patterns of incidence, clinical presentation, and treatment in the US". *Cancer* 107.7 (2006): 1448-1458.
3. Kawase K, *et al.* "Paget's disease of the breast: there is a role for breast-conserving therapy". *Annals of Surgical Oncology* 12.5 (2005): 391-397.
4. Morrogh M, *et al.* "MRI identifies otherwise occult disease in select patient with paget disease of the nipple". *Journal of the American College of Surgeons* 206.2 (2008): 316-321.
5. Lehman CD, *et al.* "MRI evaluation of the controlateral breast in women with recently diagnosed breast cancer". *New England Journal of Medicine* 365.13 (2007): 1295-1303.
6. Lundquist K, *et al.* "Intraepidermal cytokeratin 7 expression is not restricted to Paget cells but is also seen in Toker cells and Merkel cells". *American Journal of Surgical Pathology* 23.2 (1999): 212-219.
7. Marshall JK, *et al.* "Conservative management of Paget disease of the breast with radiotherapy: 10- and 15-year results". *Cancer* 97.9 (2003): 2142-2149.
8. Kawase K, *et al.* "Paget's disease of the breast: there is a role for breast-conserving therapy". *Annals of Surgical Oncology* 12.5 (2005): 391-397.
9. Siponen E, *et al.* "Surgical treatment in Paget's disease of the breast". *American Journal of Surgery* 200.2 (2010): 241-246.
10. Chen C, *et al.* "Paget disease of the breast: changing patterns of incidence, clinical presentation, and treatment in the US". *Cancer* 107.7 (2006): 1448-1457.

Volume 13 Issue 4 April 2024

©All rights reserved by Abraham Alexis Sanoh, *et al.*