
MedPeer Publisher

Abbreviated Key Title: MedPeer

ISSN : 3066-2737

homepage: <https://www.medpeerpublishers.com>

Simple Nipple Hyperkeratosis Revealing Paget's Disease

DOI: 10.70780/medpeer.000QGWD

AUTHOR AND AFFILIATION

Maha Habibi ¹, Yosra Benkraiem ¹, Nadia Ismaili ¹, Mariame Meziane ¹, Benzekri Laila¹. Department of Dermatology-Venereology, Avicenne University Hospital, Rabat, Morocco

Corresponding author: Maha Habibi .

ABSTRACT

Mammary Paget's disease is a rare condition typically associated with underlying breast carcinoma. We report the case of a 40-year-old woman presenting with a long-standing (10 years) unilateral hyperkeratotic nipple lesion without other clinical abnormalities. Dermoscopy revealed keratin clusters without vascular structures. Histopathological and immunohistochemical analysis of a punch biopsy confirmed Paget's disease (cytokeratin 7 positive). Radiological assessment, including mammography and breast MRI, showed no underlying malignancy. The patient was referred for surgical management. This case highlights an atypical presentation of Paget's disease as a chronic hyperkeratotic lesion without radiological findings, emphasizing the importance of biopsy in persistent nipple lesions to ensure early diagnosis and appropriate management.

KEYWORDS

mammary Paget's disease; nipple hyperkeratosis; dermoscopy; cytokeratin 7; breast carcinoma; case report

MAIN ARTICLE

INTRODUCTION

Mammary Paget's disease (MPD) is a rare breast malignancy involving the nipple-areola complex and is frequently associated with an underlying in situ or invasive carcinoma [1,2]. It accounts for approximately 1–4% of all breast cancers [3]. Clinically, it usually presents as an erythematous, eczematous, or ulcerative lesion of the nipple, sometimes accompanied by crusting, infiltration, or nipple retraction [1,4]. However, atypical presentations mimicking benign dermatological conditions may delay diagnosis [5].

We report an unusual case of MPD presenting as a chronic hyperkeratotic lesion of the nipple without radiological abnormalities.

CASE PRESENTATION

A 40-year-old woman with no significant medical history presented with a unilateral nipple lesion evolving over 10 years. Clinical examination revealed a well-defined hyperkeratotic plaque measuring approximately 1 cm in diameter, localized to the nipple. The lesion was non-tender, non-pruritic, and adherent to underlying tissues. No palpable breast mass or axillary lymphadenopathy was detected.



Figure 1: *Hyperkeratosis of the Nipple: Close-Up View*

Dermoscopy showed clusters of keratin without associated vascular structures.



Figure 2: *Dermoscopic Features of Nipple Hyperkeratosis: Prominent Keratin Clods*

A punch biopsy was performed. Histopathological examination revealed intraepidermal proliferation of large atypical cells with abundant pale cytoplasm and prominent nuclei, consistent with Paget cells [2]. Immunohistochemistry demonstrated strong positivity for cytokeratin 7, confirming the diagnosis of MPD [3].

Mammography and breast magnetic resonance imaging showed no evidence of underlying carcinoma, a finding reported in a minority of cases [4,6]. The patient was referred for multidisciplinary surgical management.

DISCUSSION

MPD predominantly affects postmenopausal women and is usually associated with ductal carcinoma in situ or invasive carcinoma [1,7]. Histologically, it is characterized by malignant glandular epithelial cells (Paget cells) within the epidermis, distributed singly or in clusters [2].

Clinically, MPD often mimics benign conditions such as eczema, leading to delayed diagnosis [5,8]. Symptoms include pruritus, burning sensation, and eczematous changes of the nipple-areola complex. In advanced cases, ulceration and destruction of the nipple may occur [1,4].

Diagnosis relies on histopathology supported by immunohistochemistry, with cytokeratin 7 being a highly sensitive marker [3]. Imaging studies (mammography and MRI) are essential to detect underlying carcinoma, although they may be negative in selected cases, as in ours [4,6].

Management is multidisciplinary. Surgery remains the cornerstone of treatment and includes breast-conserving surgery or mastectomy with sentinel lymph node biopsy [7,9]. Adjuvant therapies (radiotherapy, chemotherapy, endocrine therapy) depend on associated invasive disease [9].

Prognosis is mainly determined by the presence of an underlying tumor. The 10-year survival rate is approximately 90% in patients without a palpable mass but decreases significantly when invasive carcinoma is present [6,10].

This case is particularly notable due to the very long clinical evolution (10 years) and absence of radiological abnormalities. Such presentations remain uncommon and may contribute to significant diagnostic delay.

CONCLUSIONS

Although rare, MPD should be considered in any persistent nipple lesion, even in the absence of typical clinical or radiological findings. Early histological evaluation is essential to avoid diagnostic delay and ensure appropriate management.

ACKNOWLEDGEMENTS

The authors have no acknowledgements to declare and report no conflicts of interest

REFERENCES

1. Lloyd J, Flanagan AM. Mammary and extramammary Paget's disease. *J Clin Pathol*. 2000;53(10):742-749. doi:10.1136/jcp.53.10.742
<https://doi.org/10.1136/jcp.53.10.742>
2. Kanitakis J. Mammary and extramammary Paget's disease. *J Eur Acad Dermatol Venereol*. 2007;21(5):581-590. doi:10.1111/j.1468-3083.2007.02154.x
<https://doi.org/10.1111/j.1468-3083.2007.02154.x>
3. Karakas C. Paget's disease of the breast. *J Carcinog*. 2011;10:31. doi:10.4103/1477-3163.90676
<https://doi.org/10.4103/1477-3163.90676>
4. Günhan-Bilgen I, Zekioglu O, Ustun EE, et al. Paget's disease of the breast: clinical and imaging findings. *Eur J Radiol*. 2006;60(2):256-263. doi:10.1016/j.ejrad.2006.06.010
<https://doi.org/10.1016/j.ejrad.2006.06.010>
5. Sakorafas GH, Blanchard K, Sarr MG, et al. Paget's disease of the breast: a clinical perspective. *Cancer Treat Rev*. 2001;27(1):9-18. doi:10.1053/ctrv.2000.0203
<https://doi.org/10.1053/ctrv.2000.0203>
6. Dalberg K, Hellborg H, Wärnberg F. Paget's disease of the nipple in a population-based cohort. *Breast Cancer Res Treat*. 2008;111(2):313-319. doi:10.1007/s10549-007-9783-5
<https://doi.org/10.1007/s10549-007-9783-5>
7. Chen CY, Sun LM, Anderson BO. Paget disease of the breast: changing patterns of incidence and management. *J Surg Oncol*. 2006;95(2):144-149. doi:10.1002/jso.20564
<https://doi.org/10.1002/jso.20564>
8. Kothari AS, Beechey-Newman N, Hamed H, et al. Paget disease of the nipple: a multifocal manifestation of higher-risk disease. *Clin Breast Cancer*. 2002;3(3):190-195.

doi:10.3816/CBC.2002.n.024

<https://doi.org/10.3816/CBC.2002.n.024>

9. Dixon AR, Galea MH, Ellis IO, et al. Paget's disease of the nipple. Br J Surg. 1991;78(6):722-723. doi:10.1002/bjs.1800780627

<https://doi.org/10.1002/bjs.1800780627>

10. Fu W, Mittel VK, Young SC. Paget disease of the breast: analysis of 41 patients. Am J Surg. 2001;182(6):620-623. doi:10.1016/S0002-9610(01)00767-4