**SAS Journal of Surgery**
Abbreviated Key Title: SAS J Surg
ISSN 2454-5104
Journal homepage: https://www.saspublishers.com

OPEN ACCESS

**Gynaecology-Obstetrics and Endoscopy**

# Paget Disease of the Breast: An Uncommon Case Report

Benaouicha Nisrine[1*], Lazhar Hanae[1], Slaoui Aziz[1], Lakhdar Amina[1], Aziz Baydada[1]

[1]Gynaecology-Obstetrics and Endoscopy Department, Maternity Souissi, University Hospital Center IBN SINA, University Mohammed V, Rabat, Morocco

**DOI:** https://doi.org/10.36347/sasjs.2024.v10i11.007 | **Received:** 20.09.2024 | **Accepted:** 27.10.2024 | **Published:** 06.11.2024

***Corresponding author:** Benaouicha Nisrine
Gynaecology-Obstetrics and Endoscopy Department, Maternity Souissi, University Hospital Center IBN SINA, University Mohammed V, Rabat, Morocco

**Abstract** **Case Report**

Paget's breast disease is a rare malignancy with an incidence of 0.7% to 4.3% among all primary breast cancer. It's characterized by an eczemous and ulcerative lesion of the nipple area. It's associated up to 90% to an underlying invasive carcinoma or carcinoma in situ. Upon every cutaneous lesion matching the description above in women in their fifth or sixth decade, a punch biopsy should be performed. Imaging investigations rely on mammography, breast ultrasound and most importantly on Magnetic resolution imaging (MRI). Treatment management offers a wide array of options ranging from total mastectomy with sentinel node biopsy to non-surgical and non-invasive therapy. We present the uncommon case of a 49 years old female presented to our facility with Paget breast disease treated by breast conserving surgery.

**Keywords:** Paget's disease, Breast cancer, Invasive ductal carcinoma, Nipple lesion, Breast conservation surgery.



## INTRODUCTION

Paget's Breast Disease is a scarce malignancy. It's majorly associated with underlying ductal carcinoma in situ or invasive breast cancer. Clinically, it presents itself commonly between 50 and 60 years old females as an eczematoid, itchy erythema or ulceration located in the nipple-areolar area that may be associated with nipple discharge or even nipple retraction. We hereby present the uncommon case of a Paget breast disease associated with ductal invasive carcinoma in a 49 years old female patient.

## CASE PRESENTATION

A 49 years old woman was referred by her primary physician to our out-patient gynecology department of the national institute of oncology in order to further investigate an eczematous lesion of the left nipple area. The patient's history revealed a perimenopausal state, 3 vaginal deliveries, no previous mammography, no pap test and no family or personal history of cancer. Her primary complaint was pruritis of the left nipple area that have been lingering for the last 6 months followed by the surfacing of an eczematous miscolored lesion with a dark beading (Image 1).

**Image 1: Eczematous squamous miscolored lesion of the nipple area circled by a dark beading**

Clinical examination showed no palpable breast mass and no axillary lymph nodes. The patient underwent a punch biopsy of the lesion that concluded on Paget's disease invading the epidermis.

Upon these findings, further imaging investigations were led in order to single out the underlying carcinoma. Mammography followed by an ultrasound were the initial radiological investigations that turned out to be negative. Hence, an MRI was order showing an enhancing irregular mass in the outer -upper quadrant of the left breast with focal enhancement along the nipple areolar region. Ergo, the patient underwent an MRI guided breast micro biopsy. The histopathology findings declared an invasive ductal carcinoma of the breast with estrogen receptor positive (ER+), progesterone receptor positive (PR+), and human epidermal growth factor receptor 2 positive (HER2+).

Based on the diagnosis of Paget's disease of the breast associated with an invasive ductal carcinoma the patient was presented two options: a lumpectomy with a sentinel node biopsy or a mastectomy also with a sentinel node biopsy.

She opted for a lumpectomy which was performed with no complication. Surgical pathology revealed invasive ductal carcinoma with clear margins and nipple involvement of Paget's disease of the breast. The findings of the sentinel lymph node biopsy were negative for metastasis.

Post-operative care was eventful and the patient is scheduled for her follow up in 2 months.

## DISCUSSION

First described in 1874 by James Paget, Paget's disease of the breast (PDB) is a scarce breast malignancy where its most clinical manifestation is a skin lesion located in the nipple-areolar complex (NAC) [1]. Epidemiologically, PBD accounts for 0,7% - 4,3% among all primary breast cancer [2].

Histopathologically, it's distinguished by the presence of Paget cells infiltration with abundant pale cytoplasm and nuclei located centrally in the epidermal layer of the NAC [3].

We can divide PDB in three categories: PDB with invasive ductal carcinoma, PDB with underlying ductal carcinoma in situ and PDB alone. Our report sheds the light on a case of PDB with invasive ductal carcinoma [4].

Pathogenesis of PDB Is a subject of immense controversy. Some epidermotropic authors speculate that the migrating ductal cancer cells along the basal membrane of the nipple are the origin of Paget's cells whereas others postulate that they originate from malignant transformation of keratinocytes giving rise to PDB [5-7].

The initial NAC lesion evolves gradually over months to years. Symptoms associated are usually pruritus, redness, discharge, oozing and scaling [8, 9]. Physical examination typically finds a sharply delimited, fine scaly, erythematous, crusty and thickened plaques on the nipple accompanied by skin ulceration and nipple retraction in advanced stages [10]. It's important to underline the possibility of PDB arising from the axillary accessory breast tissue or the supernumerary nipples [11].

Since PDB coexists with breast carcinoma - invasive or insitu- in more than 90% of cases, imaging is the stepping stone to rule out or single out malignancy. Mammography is the first radiological investigation tool with a 97% sensitivity in detecting an underlying malignancy in PDB if there is a clinically palpable mass and a specificity of 50% if no mass is palpable[12-13]. If its findings are negative, an ultrasound may be performed. No palpable mass alongside negative mammography and ultrasound are the true indication for breast MRI. MRI findings include NAC thickening and enhancement, detecting an underlying mass, enhancing ductal carcinoma in situ or a combination of these aspects [14]. Furthermore, in case of positive mammography and ultrasound, MRI helps assess the extent of the disease especially when breast-conserving surgery is contemplated.

Despite imaging findings, every cutaneous lesion of the NAC associated with a rash should be subject to a punch biopsy so as to infirm or affirm a PDB diagnosis [15]. Immuhistochemistry is a powerful tool in detecting the molecular categories of PDB and differentiating PDB from other skin lesions [16].

Upon diagnosis, neoplasic staging isn't altered by the presence of PDB. If PDB isn't associated with invasive or insitu carcinoma, it's classified as Tis (Paget) disease.

Treatment options include surgical management, radiotherapy, photodynamic therapy or even endocrine therapy. The surgical approach consists of breast- conserving therapy in cases of unifocal disease limited to the NAC and of total or skin-sparing mastectomy with surgical axillary staging in cases of multicentric or multifocal disease [14]. Thanks to oncoplastic surgical techniques, to NAC reconstruction and to dermatography, aesthetic outcomes have been very pleasing [15].

## CONCLUSION

Paget's disease of the breast is a rare malignancy associated majorly to an underlying invasive or in situ cancer. Suspicion should rise subsequently to a persistent skin lesion located in the nipple-areolar

complex characterized by itchiness, eczematoid aspect, soreness and redness. Though Paget's disease is predominantly a clinical diagnosis, negative mammography cannot reliably exclude underlying malignancy. Both clinical and imaging findings are complementary to each in order to make a diagnosis.

## REFERENCES


1. Sakorafas, G. H., Blanchard, K. D., Sarr, M. G., & Farley, D. R. (2001). Paget's disease of the breast: a clinical perspective. *Langenbeck's archives of surgery*, *386*, 444-450.
2. Kanitakis, J. (2007). Mammary and extramammary Paget's disease. *Journal of the European Academy of Dermatology and Venereology*, *21*(5), 581-590. doi:10.1111/j.1468-3083.2007.02154.x
3. Wong, S. M., Freedman, R. A., Sagara, Y., Stamell, E. F., Desantis, S. D., Barry, W. T., & Golshan, M. (2015). The effect of Paget disease on axillary lymph node metastases and survival in invasive ductal carcinoma. *Cancer*, *121*(24), 4333-4340. doi:10.1002/cncr.29687
4. Chen, C. Y., Sun, L. M., & Anderson, B. O. (2006). Paget disease of the breast: changing patterns of incidence, clinical presentation, and treat- ment in the U.S. *Cancer, 107*(7), 1448-1458. doi:10.1002/cncr.22137
5. Markarian, S., & Holmes, D. R. (2022). Mammary Paget's Disease: An Update. *Cancers (Basel)*, *14*(10), 2422. https://doi.org/10.3390/cancers14102422
6. Di Tommaso, L., Franchi, G., Destro, A., Broglia, F., Minuti, F., Rahal, D., & Roncalli, M. (2008). Toker cells of the breast. Morphological and immunohistochemical characterization of 40 cases. *Human pathology*, *39*(9), 1295-1300. https://doi.org/10.1016/j.humpath.2008.01.018
7. Lopes Filho, L. L., Lopes, I. M., Lopes, L. R., Enokihara, M. M., Michalany, A. O., & Matsunaga, N. (2015). Mammary and extramammary Paget's disease. *An Bras Dermatol*, *90*(2), 225-231. https://doi.org/10.1590/abd1806-4841.20153189
8. Yasir, M., Khan, M., & Lotfollahzadeh, S. (2022). Mammary Paget disease. StatPearls [Internet]. StatPearls Publishing, Treasure Island, Fl; 2022.
9. Adams, S. J., & Kanthan, R. (2016). Paget's disease of the male breast in the 21st century: A systematic review. *The Breast*, *29*, 14-23. doi:10.1016/j.breast.2016.06.015
10. Salati, S. A., Alsulaim, L., & Rather, A. A. (2022). Paget's Disease of the Breast (PDB) – A Review. *Journal of Analytical Oncology*, 11, 45–53. https://doi.org/10.30683/1927-7229.2022.11.07
11. El Khoury, M., Lalonde, L., David, J., Issa-Chergui, B., Peloquin, L., & Trop, I. (2011). Paget's disease of the axilla arising from an underlying accessory mammary tissue. *Clinical radiology*, *66*(6), 575-577. https://doi.org/10.1016/j.crad.2010.12.010
12. Lim, H. S., Jeong, S. J., Lee, J. S., Park, M. H., Kim, J. W., Shin, S. S., ... & Kang, H. K. (2011). Paget disease of the breast: mammographic, US, and MR imaging findings with pathologic correlation. *Radiographics*, *31*(7), 1973-1987. https://doi.org/10.1148/rg.317115070
13. Kothari, A. S., Beechey-Newman, N., Hamed, H., Fentiman, I. S., D'Arrigo, C., Hanby, A. M., & Ryder, K. (2002). Paget disease of the nipple: a multifocal manifestation of higher-risk disease. *Cancer*, *95*(1), 1-7. https://doi.org/10.1002/cncr.10638
14. Sripathi, S., Ayachit, A., Kadavigere, R., Kumar, S., Eleti, A., & Sraj, A. (2015). Spectrum of imaging findings in Paget's disease of the breast—a pictorial review. *Insights into Imaging*, *6*(4), 419-429. https://doi.org/10.1007/s13244-015-0415-z
15. Markarian, S., & Holmes, D. R. (2022). Mammary Paget's disease: an update. *Cancers*, *14*(10), 2422. https://doi.org/10.3390/cancers14102422
16. Barnes, P. J., Dumont, R. J., & Higgins, H. G. (2007). Acinar pattern of mammary Paget's disease: a case report. *The Breast Journal*, *13*(5), 520-526. doi:10.1111/j.15244741.2007.00477.x