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Paget's Disease of the Nipple: A Case Report and Review of the Literature

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Abstract

Paget's disease of the breast is a particular form of the disease that occurs in 1 to 4% of breast cancers. It is characterised by an accumulation of abnormal cells known as "PAGET cells" in the epidermis of the nipple, associated with underlying glandular cancer in 82 to 100% of cases **[1]**. Clinically, it presents as eczematous eruptions on the nipple and areola, often unilateral. Imaging based on ultrasound mammography and magnetic resonance imaging of the breast is therefore useful in searching for underlying cancer. Its management and prognosis are conditioned by the accompanying breast cancer.

Based on an observation and a review of the literature, this article sets out to explain the clinical features and therapeutic management of this disease.

Keywords: Paget's Disease of the Breast, Case Report, Invasive Cancer, Carcinoma in Situ, Surgery, Sentinel Lymph Node

Identity

The patient was 60 years old, naïve, with no previous pathological history, and presented with an eczematous lesion of the right nipple. The lesion had begun a year earlier as a scaly, pruritic patch involving the nipple and areola of the right breast. Clinical examination revealed an inflammatory plaque on the right nipple, ulcerated in places and topped with a crust **(Figure 1)**. The rest of the examination of the right breast was unremarkable, in particular there was no palpable nodule or nipple discharge. The left breast was unremarkable and the axillary folds were free.

Mammography revealed a pseudo-nodular, water-toned opacity in the nipple area, with imprecise boundaries and segmental micro-calcifications; the rest of the mammogram was unremarkable.

Ultrasound examination of the breast showed a pseudo-nodule measuring 27 mm in length, located in the right retroareolar region, poorly defined, hypoechoic, heterogeneous, with posterior attenuation. The rest of the examination was unremarkable.

The echomammographic examination was classified as BIRADS 5 on the right and 1 on the left **(Figure 2,3)**.

A biopsy was performed after ultrasound detection, as well as a nipple biopsy. The results showed a high-grade intracanal carcinoma with the presence of a microfocus measuring less than 0.1 cm long of an infiltrating carcinoma of type NOS grade II of SBR, with Paget's disease of the nipple, Immunohistochemistry was not performed.

The extension work-up included abdominal ultrasound, chest X-ray, bone scan and CA15-3 assay, all of which were negative.

The patient underwent lumpectomy with removal of the nipple-areolar plate (pamectomy) and a sentinel lymph node **(Figure 4)**.

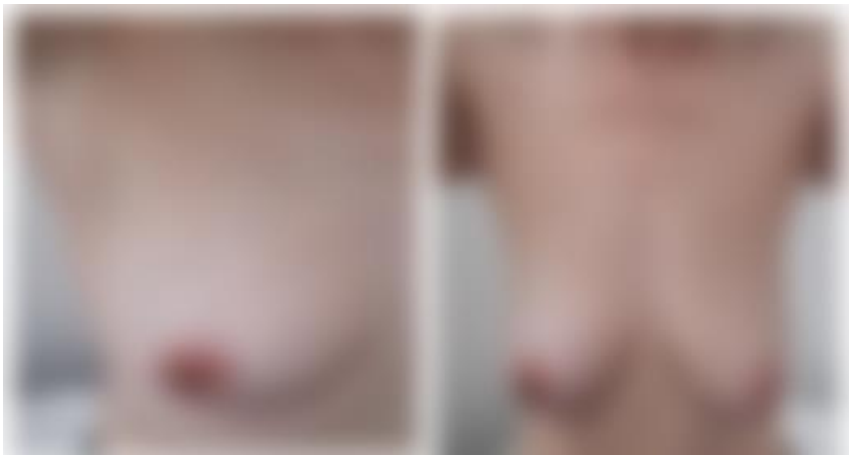

Fig 1. On the right breast, there was an inflammatory placard on the nipple, ulcerated in places and topped with a crust.

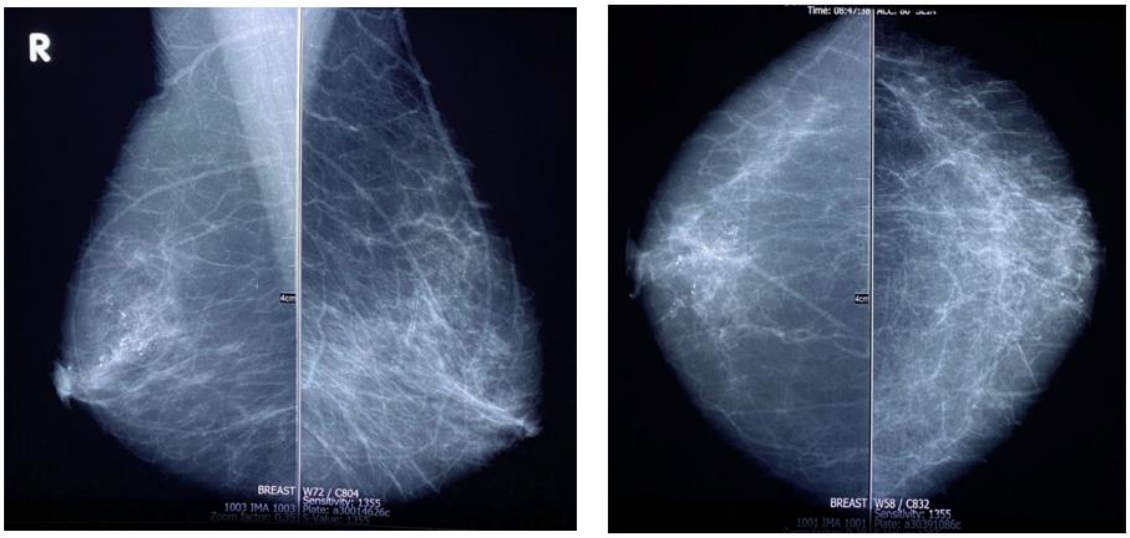

Fig 2: Surcroît d'opacité pseudo-nodulaire, de tonalité hydrique rétro-mamelonnaire à limites imprécises comportant des micro-calcifications segmentaires. Mammographie [(A) – incidence de profil; (B) – incidence de face].

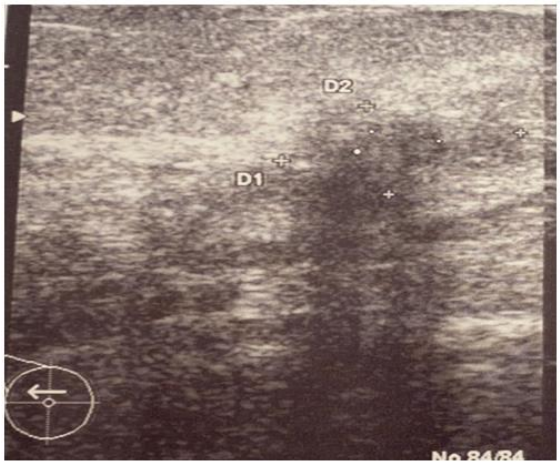

Fig 3: Poorly limited, hypoechoic, heterogeneous pseudo nodule, with posterior attenuation, measuring 27 mm in long axis.

Fig 4: Surgical specimen of the right lumpectomy with removal of the areola-mammelon plate.

The final anatomopathological examination revealed the presence on the oriented pamectomy specimen of a cutaneous-mammary tumour, measuring 3.5 cm in length and occupying the entire mammelon. It was located 5 mm from the nearest internal excision site and corresponded to a pure intracanal carcinoma, of solid and cystic type, associated with significant carcinomatous pagetoid migration with excision sites in healthy areas, with no invasive component on the specimen **(Figure 5,6)**.

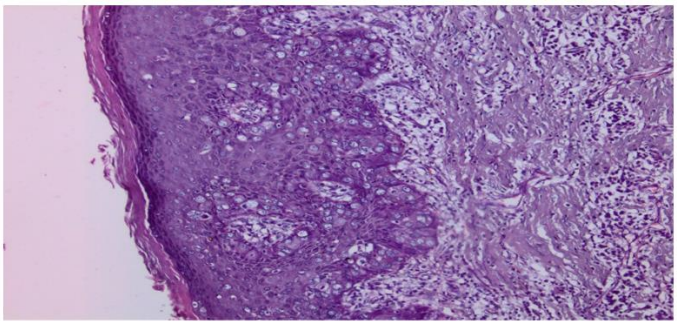

Fig 5: Intraepithelial tumour proliferation arranged in clusters within orthokeratotic squamous epithelium (under standard staining, x200).

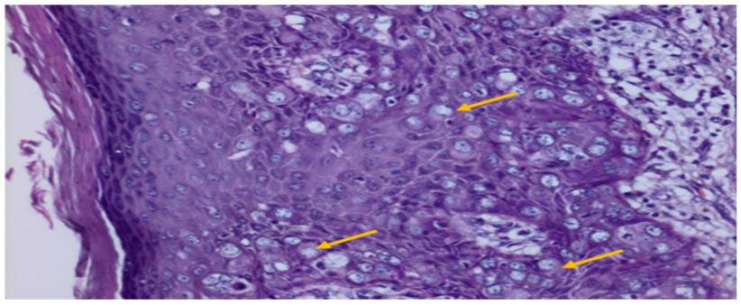

Fig 6: Tumour cells [orange arrows] have abundant pale cytoplasm and large, irregularly outlined nuclei with a single nucleolus (under standard staining, x400).

The patient was referred to the oncology department for further management.

Discussion

Paget's disease was first described by Sir Paget in 1874 as an eczematous lesion of the nipple associated with an underlying cancer **[2]**. It is a rare disease, accounting for less than 5% of breast cancers **[3]**. It mainly affects post-menopausal women, with an average age of 62.6 years. No clinical or epidemiological predisposing factors have been described **[4]**.

Paget's disease of the breast may be associated with homolateral breast cancer in 82 to 100% of cases, including 13.3 to 52% with carcinoma in situ and 30 to 60% invasive, or it may be isolated, including 1.4 to 13.3% **[5]**.

In cases where carcinoma in situ is associated, multifocality is described in 42 to 63% of cases.

Paget's disease is explained by several theories, including two main ones:

- The 1st theory maintains that luminal lactiferous duct epithelial cells give rise to Paget's cells, which migrate retrogradely into the overlying epidermis.
- The second theory, independent of the underlying cancer, suggests that Paget's cells transform in situ and derive from cells of the terminal lactiferous duct at its junction with the epidermis. This may explain situations in which PD is not associated with an underlying carcinoma or is anatomically distant from it **[6,7]**.

Diagnosis is often delayed, with constant involvement of the nipple in the initial stage, notably with the presence of pruritus and a nipple that becomes red and shiny; in the intermediate stage, the nipple becomes thick, reddened and scaly; and in the late stage, erosion and indurated crusting with clear boundaries appear, extending very slowly and centrifugally from the nipple towards the areola. Bloody or serous discharge from the nipple is present in 30-60% of cases, with a palpable mass which may be distant from the nipple in 30-50% of cases **[8]**.

In the presence of an eczematous lesion, the main differential diagnosis of Paget's disease is nipple eczema, but the unilateral nature, progressive nature and lack of response to corticosteroid therapy make it possible to diagnose the disease response to corticosteroid therapy. Other differential diagnoses, such as psoriasis, superficial basal cell carcinoma and melanoma, may also be considered, but only histology can confirm the diagnosis.

Whenever Paget's disease is suspected, mammography is performed to detect heterogeneous, poorly defined microcalcifications with suspicious opacities. The sensitivity of mammography to detect a tumour is 97% in the presence of a palpable mass, whereas it is only 50% in the absence of a palpable mass **[8]**. Breast ultrasound is routinely performed to find formations that attenuate ultrasound and to aid biopsy. Additional specialised imaging techniques such as MRI can be used to create further images of the breast and to determine whether underlying cancer is present **[3,9]**.

Paget's disease is diagnosed with certainty either by cytological scraping of the nipple or, at best, by nipple-areolar biopsy. Histologically, this lesion is characterised by the presence of pagétic cells in the epidermis of the nipple. These are large cells with clear cytoplasm and a large irregular hyperchromatic nucleus, the site of mitosis **[10]**. It is important to biopsy not only the superficial lesion, but also any underlying abnormalities detected on physical examination or imaging.

Radical treatment (mastectomy) has long been considered the appropriate treatment for Paget's disease of the nipple, due to its frequent association with multifocal or multicentric breast cancer. In their study of 70 patients with Paget's disease of the nipple, Kothari et al. found that 60% had invasive carcinoma, more than a third of which was palpable **[5]**. 41% of this group had multifocal breast cancer and 30% multicentric. In the group with underlying non-palpable breast cancer, multifocality and multicentricity were found in 60% of cases. Currently, the majority of teams adopt conservative treatment. This treatment consists of performing a central lumpectomy removing the nipple-areolar plate, followed by radiotherapy **[4,11]**, even if there is a limited associated cancer proven by imaging **[12]**.

The evaluation of lymph node involvement (sentinel lymph node technique) in cases of Paget's disease of the nipple is not yet clear. This would be considered in cases of association with radiologically and histologically proven invasive cancer **[13]**.

Currently no data are available to support the use of endocrine therapy in the MPD without an associated DCIS or invasive carcinoma **[3]**.

Post-treatment surveillance is correlated with that of the breast carcinoma with which it is associated, and the prognosis of Paget's disease of the nipple is conditioned by the association with an underlying cancer **[12]**.

Conclusion

Paget's disease of the nipple is a rare entity that should be suspected in the presence of any eczematous lesion of the nipple that is persistent and resistant to medical treatment.

Diagnostic certainty requires histological confirmation. The combination of mammography and breast ultrasound,

possibly with breast MRI, should look for.

Conservative treatment of Paget's disease of the nipple is a defensible alternative in patients with limited associated cancer, proven by imaging. The prognosis of Paget's disease of the nipple is determined by the invasiveness of the underlying cancer.

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