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Breast Paget Disease: A Rare Cancer in Dermatology

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Abstract **Case Report**

Paget's disease (PD) of the breast is a rare disease of the nipple-areola complex often associated with underlying carcinoma, accounting for less than 4% of all breast carcinomas. We report a case of Paget's disease remarkable for its rarity, the diagnostic delay and the relatively young age of our patient in the Dermatology department of the Donka National Hospital. A 38-year-old woman consulted for itching of the left nipple associated with an oozing erythematous-squamous plaque and tumor-like left axillary lymphadenopathy evolving since4 years. Clinical examination and histopathology confirmed breast Paget's disease. A left mastectomy with lymph node dissection was performed, supplemented by chemotherapy. The evolution was good with absence of recurrence and metastasis after a follow-up of 16 months. Paget's disease corresponds to the invasion of the nipple epidermis by ductal carcinoma in situ (CIS), a rare variant of CIS. Classically, it develops between the ages of 51 and 70, however cases of Paget's disease of the nipple in young women during periods of genital activity have been reported in the literature as illustrated by our observation. The diagnosis of PD is often made late. An easy biopsy should be offered for all unilateral eczematous lesions of the nipple to compensate for this diagnostic delay. Mastectomy or breast-conserving surgery is often recommended. Adjuvant treatments are discussed on a case-by-case basis.

Keywords: Paget's disease, Young age, Dermatology.

CONTEXT

Paget's disease (PD) of the breast is an infiltration of the epidermis of the nipple by adenocarcinoma-like cells [1]. This rare breast neoplasia varies between 0.5 to 4% of all breast carcinomas and remains associated with ipsilateral breast cancer in 82 to 100% of cases [2]. It is secondary either to the in situ transformation of squamous cells (Toker cells); either to the migration at the level of the aero-mammary plaque of the underlying carcinomatous cells [3]. The diagnosis is often made late in relation to the insidious onset of breast symptoms and is based on a skin biopsy [4]. We report a case of Paget's disease remarkable for its rarity, the diagnostic delay and the relatively young age of our patient in the Dermatology department of the Donka National Hospital.

OBSERVATION

This was a 38-year-old married patient with no particular pathological history, who was seen for a dermatological consultation for pruritic erosion of the left nipple that had been present for 4 years. This lesion began as a small, finely scaly erythematous lesion on the nipple, very itchy. The physical examination revealed an eythematous-squamous plaque of the left nipple, occasionally oozing measuring 3.5 cm in long axis, associated with an intra-mammary nodule, poorly defined ipsilateral and mobile in relation to the cutaneous plane (Figure 1). Palpation of the axillary hollows revealed tumor-like lymphadenopathy. The remainder of the physical examination was unremarkable. The diagnostic hypotheses mentioned are: Paget's disease of the breast, Contact eczema, Erosive adenomatosis of the nipple. The histology of the nipple skin biopsy showed a proliferation of large cells with clear, basophilic cytoplasm, with an eccentric nucleus with a “chaton ring”. The cells are dispersed within the epithelium with in some places a clear pagetoid migration (HEx100) thus confirming Paget's disease (Figure 2). Ultrasound mammography found, at the retroaerolar level, microcalcifications and infiltration of the opposite

subcutaneous fat measuring 20x10 mm in diameter, associated with ipsilateral axillary lymphadenopathy, classified ACR V (Figures 3 and 4). The patient underwent a left mastectomy with lymph node dissection. The definitive histological analysis concluded that there was an infiltrating ductal carcinoma with Paget's disease of the nipple and lymph node invasion. The patient received chemotherapy. The evolution was good with absence of recurrence and metastasis after a follow-up of 16 months.

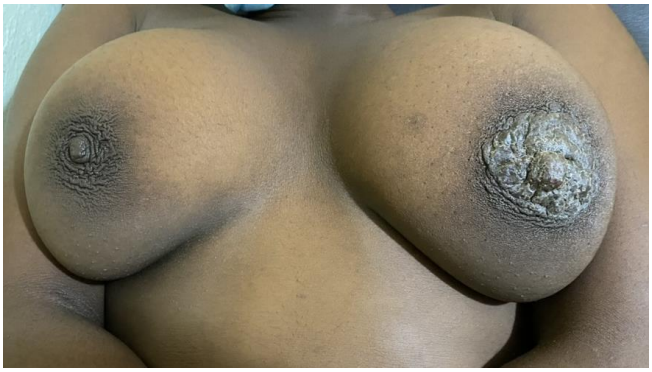

Figure 1: Erosion of the left nipple covered by crusts

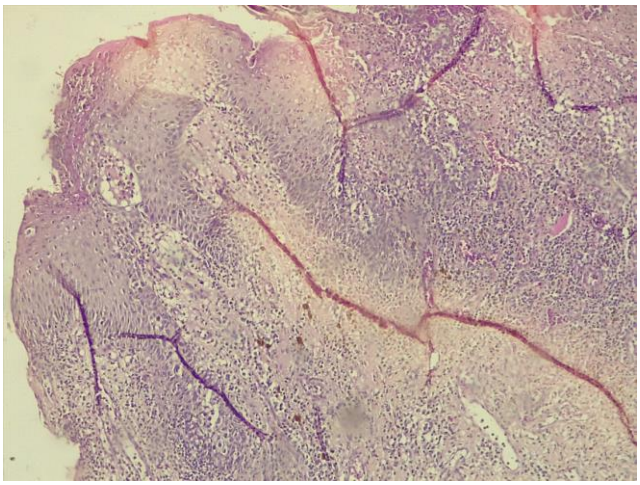

Figure 2: Pagetoid infiltration of the epidermis by atypical cells (HE x100)

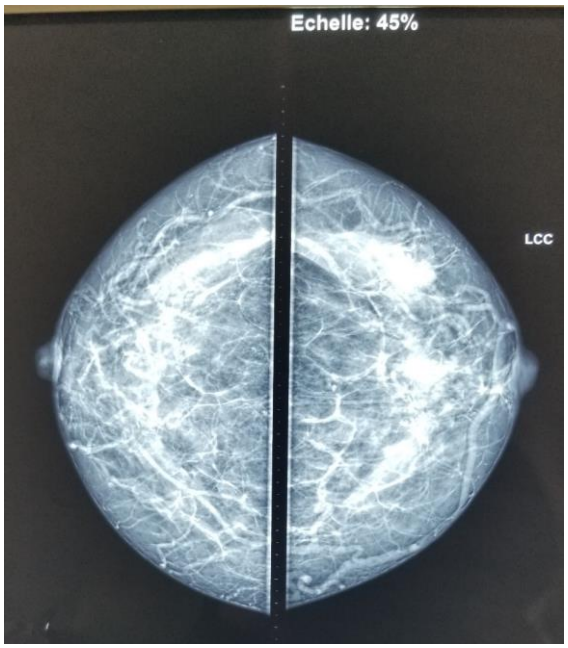

Figure 3: Contours speculated with microcalcifications and fat infiltration retroaerolar

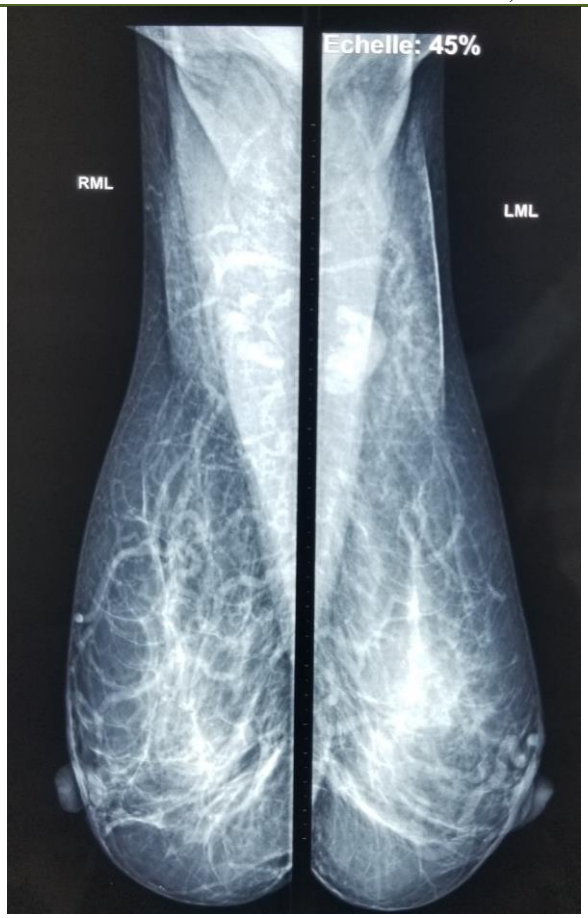

Figure 4: Infiltration of retroaerolar subcutaneous fat measuring 20x10mm associated with ipsilateral axillary lymphadenopathy

DISCUSSION

PD was first described in 1874 by Sir James Paget, a British surgeon and physiologist [4]. It is secondary to the degeneration of nipple keratinocytes. However, the most accepted theory is the invasion of the epidermis of the nipple by paget cells originating from ductal carcinoma in situ (CIS). The multifocality of ductal carcinomas in situ, associated with Paget's disease is found between 42 to 63% of cases [4–6]. It frequently occurs in postmenopausal women after the sixth decade of life, however it has also been diagnosed in young adults during the period of genital activity as in our patient. No epidemiological or clinical factor is recognized as predisposing to the occurrence and development of this neoplasia [1, 7].

PD must be suspected and actively sought in the face of any unilateral modification of the nipple and/or areola of the breast, often associated with functional signs (pruritus or pain). Its onset is insidious, evolving over several months or even years. It begins in the nipple, then extends to the areola, and in some advanced cases to the surrounding skin, in the form of eczematous, erythematous, thickened, moist or crusted lesions with irregular edges, with or without fine desquamation, nipple induration, discharge, bleeding, ulceration and intussusception [4, 5, 8]. Our patient presented with a pruritic erythematous-scaly plaque on the left nipple with an insidious onset and a chronic course.

The delay in diagnosing PD in the majority of patients could be explained by the appearance similar to common skin pathologies such as those suggestive of eczema, psoriasis but above all linked to the reluctance of patients to accept such a minimal lesion as cancerous. in an apparently healthy woman without a palpable breast mass [4, 5, 9]. In our patient, the diagnostic delay was linked to the reluctance to accept the hypotheses raised at the start of follow-up, non-compliance with prescribed follow-up appointments but also by the use of antihistamine for self-medication. Such a diagnostic delay could be avoided by assuming that any unilateral itchy rash on the nipple-areola complex is Paget's disease until proven otherwise but also by interviewing our patients upon first contact [4].

The definitive diagnosis of PD is made either by cytological scraping of the nipple and/or by skin biopsy on the nipple-areola complex. Histologically, pagetoid cells are found in the epidermis of the nipple. These are large cells with clear cytoplasm and a large irregular hyperchomatic nucleus, the site of mitosis [8]. In our patient, with hematoxylin-eosin staining, a pagetoid infiltration of the epidermis by large cells with clear

cytoplasm, irregular contours, hyperchromatic and dyskaryotic, grouped in clusters was revealed, thus confirming Paget's disease breast.

Radiological exploration is necessary for the active search for underlying breast cancer in order to guide management. The radiological abnormalities found can be variable. Mammography can detect masses or calcifications that are related to invasive or in situ ductal carcinoma, particularly when these lesions are palpable. However, a negative examination does not exclude the presence of tumors. Breast ultrasound is particularly useful in cases where mammograms are negative or contraindicated. However, the findings are nonspecific, showing only parenchymal heterogeneity, hypoechoic areas, discrete masses, skin thickening, and ductal dilation. Magnetic resonance is very sensitive for diagnosing breast tumors, especially if mammography and ultrasound are normal. It makes it possible to demonstrate a thickening of the papillary-areolar complex, an increase in the size of the nipple, the detection of ductal lesions in situ and invasive tumors, even if clinically absent [4, 10, 11]. In our patient, echomammography found, at the retroaerolar level, microcalcifications and infiltration of subcutaneous fat measuring 20x10 mm in diameter, associated with ipsilateral axillary lymphadenopathy, thus confirming the clinical hypotheses mentioned.

Treatment essentially depends on whether or not there is underlying breast cancer. It consists of performing a total mastectomy or breast-conserving surgery with lymph node dissection. Total mastectomy has long been considered the appropriate treatment for PD of the nipple, due to its association with multifocal or multicentric breast cancer. When the diagnosis of breast cancer is established early, conservative surgery constitutes a plausible alternative. It consists of performing a central lumpectomy removing the nipple-areola plaque followed by radiotherapy. Adjuvant treatments (radiotherapy, chemotherapy, immunotherapy and hormonal therapy) are initiated on a case-by-case basis [1, 3–5]. Given the clinical picture presented by our patient, combining a palpable breast mass and ipsilateral axillary lymphadenopathy; a radical mastectomy was performed combined with chemotherapy in the oncological surgical unit. Certain factors indicate an unfavorable prognosis, including: the presence of a palpable breast tumor, enlarged lymph nodes, histological type of breast cancer and age less than 60 years [2, 4]. After a 16-month follow-up, our patient had a good clinical progress without signs of recurrence or metastases and respected her prescribed follow-up appointments at the Oncological Surgery Unit.

CONCLUSION

Breast Paget's disease is rarely reported. The skin remains the mirror of the body. This breast carcinoma occurred in a patient during a period of genital activity in our context, marked by a delay in treatment and a favorable surgical outcome.

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