Case Report

Mammary Paget's Disease in a 54-year-old Filipino Female

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Abstract

Mammary Paget's disease (MPD) is a rare form of intraepithelial adenocarcinoma occurring in the apocrine gland-bearing areas in patients older than 50 years old. This clinical disease presents as erythematous, scaly plaque that usually affects the unilateral nipple or areola and is frequently misdiagnosed as inflammatory or infectious dermatitis. In this report, we are presented with a 54-year-old Filipino female who came in with a 3-year history of persistent pruritic erythematous moist plaque on the right nipple gradually spreading to the surrounding areola previously treated as a case of fungal infection. Mammography revealed BI-RADS 4C. Skin punch biopsy showed nuclear atypia with pale staining cytoplasm and were consistent with MPD. Immunohistochemical stains showed positive for epithelial membrane antigen and carcinoembryonic antigen. We reiterate the importance of early diagnosis for appropriate treatment to prevent unwanted sequelae.

Keywords: Mammary Paget's disease, MPD, Paget's disease of the breast

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Introduction

Mammary Paget's disease (MPD) or Paget's disease of the breast is a rare form of intraepithelial adenocarcinoma of the nipple and/or areola.[1] It was approximately 0.7%–4.3% of all breast cancers[2] and is seen in <1% of women.[3] A national study in China analyzing data from 2016 reported a prevalence of MPD at 0.42 per 100,000 population, with a marked female predominance.[2] This was originally described by Sir James Paget in 1874.[4] This report aims to highlight the clinical presentation, diagnosis, and management of MPD. This clinical entity is characterized by chronic nipple eczema, bleeding, or development of a mass.[5,6] Other associated symptoms include pruritus and may be associated with ulceration, weeping, and crusting.[5,6] It is crucial to distinguish Paget disease of the breast from other similar conditions such as inflammatory skin disorders, precancerous skin conditions, and breast tumors.[5] By discussing the case of a 54-year-old patient diagnosed with MPD, the report seeks to emphasize the importance of accurate diagnosis, the challenges in distinguishing MPD from other conditions, and the successful treatment outcomes with mastectomy and radiotherapy. In addition, it aims to raise the awareness of this uncommon disease, particularly in the context of limited epidemiological data from the third world countries.

Case Report

A 54-year-old Filipino woman presented with a 3-year history of persistent pruritic erythematous plaque on the right nipple gradually spreading to the surrounding

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areola. She was diagnosed with fungal infection upon several consultations with a private physician; however, she remained unresponsive to topical antifungal after a year. The patient had a history of right oophorectomy (2016) and total abdominal hysterectomy with bilateral salpingo-oophorectomy (2018) due to endometriosis, renal transplantation (2018), and unrecalled bladder surgery (2018). Physical examination revealed a solitary, well-defined, irregularly-shaped erythematous, moist plaque measuring 11 cm × 12 cm with hyperpigmented well-demarcated border located on the right breast [Figure 1]. The patient has no history of fever, weight loss, and malaise. No palpable breast mass was noted with no cervical, hilar, and axillary lymphadenopathy. The rest of the physical examination was normal.

A 4-mm skin punch biopsy was done from the right areola. The microsections showed cells with nuclear atypia and pale staining cytoplasm all throughout the epidermis while the dermis revealed superficial and mid-perivascular infiltrates of lymphocytes, histiocytes, and plasma cells [Figure 2a]. The immunohistochemical studies revealed positive for epithelial membrane antigen (EMA) [Figure 2b] and carcinoembryonic antigen (CEA) [Figure 2c]. These findings were consistent with MPD. Hematologic and biochemical tests were unremarkable. Gynecologic ultrasound was normal. Breast ultrasound revealed an ill-defined echogenic focus right peri-areolar on the 8 o'clock region signed of as probable inflammatory disease. Mammogram showed irregular hyper-dense lesion and regional punctate calcifications on the right breast with a score of BI-RADS 4C which translated to a moderate suspicion of malignancy, having a 50%–90% likelihood of malignancy. There was no family history of malignancy. The patient was referred to Surgery and Oncology service wherein modified radical mastectomy of the right breast followed by breast radiotherapy was done. At present, all lesions have resolved after 1 year of radiotherapy.

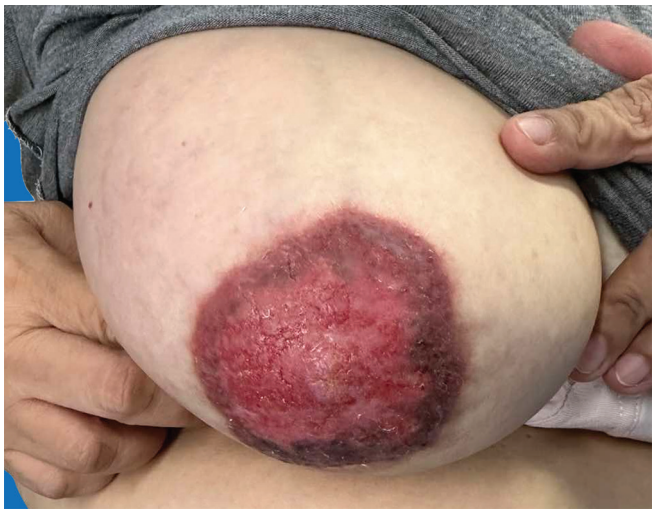

Figure 1: Solitary, well-defined, irregularly-shaped erythematous moist plaque measuring 11 cm × 12 cm with hyperpigmented well-demarcated border on the right breast

Discussion

MPD, which was first described in 1874,[4] is characterized by eczematous changes in the nipple-areolar complex and commonly associated with an underlying *in situ* or invasive ductal carcinoma.[5] It is usually localized and unilateral. Advanced cases may show extension of the lesion to the surrounding skin.[7] Two theories have been proposed to explain the pathogenesis of this disease. The most accepted theory is the epidermotropic theory which suggests that Paget's cells arise from the ductal cells that had migrated from the underlying breast parenchyma.[8] The clinical characteristics of MPD are nonspecific and resemble those of various inflammatory, reactive, and neoplastic conditions.[7]

In the case presented, the clinical presentation of a chronic erythematous moist plaque on the unilateral nipple together with the age and gender of the patient brought us to an initial diagnosis of MPD despite patient

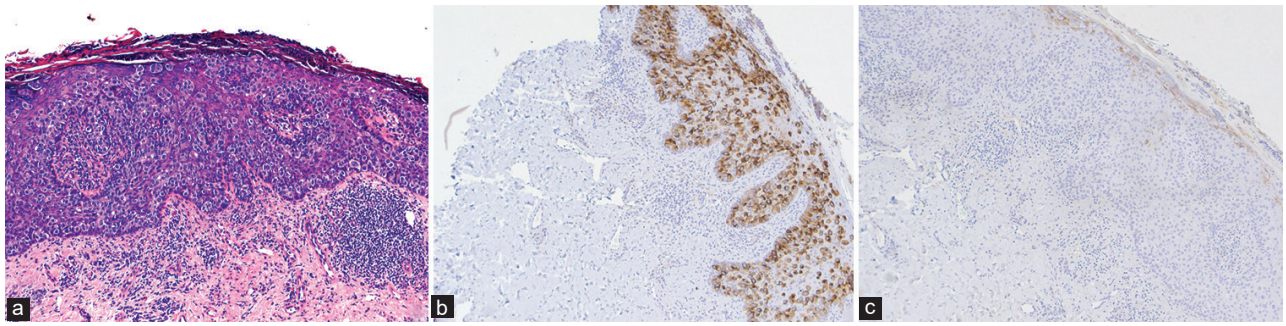

Figure 2: Hematoxylin and Eosin stain of skin punch biopsy specimen obtained from the right areola showing cells exhibiting nuclear atypia and pale cytoplasm all throughout the epidermis, 100x (a), Epithelial membrane antigen stain showing a positive result, 100x (b), Carcinoembryonic Antigen stain showing a positive result 40x (c)

being misdiagnosed as a case of fungal infection. This underscores the diagnostic challenge of MPD, which frequently mimics benign inflammatory dermatoses. Studies have reported that the time interval between the initial appearance of symptoms and the correct diagnosis of MPD can range from 6 months to over a year, with a mean delay of approximately 8 months in some cohorts. In certain series, up to 40% of patients experienced a diagnostic delay exceeding 12 months. This lag in the diagnosis may permit the progression of underlying ductal carcinoma, thus impacting treatment complexity and patient prognosis. A size ranging from 3 mm to 15 cm in diameter may present with underlying malignancy in up to 88% of the cases.[8] Fifty percent of the cases is associated with a palpable mass. In our case, no mass was palpated but with a high index of suspicion, we worked up the patient and mammography showed BI-RADS 4C which confirmed our diagnosis. This reinforces the importance of imaging and histopathological assessment even in the absence of clinical mass. According to Yasir *et al*. (2024), 20% of patients may show the presence of abnormality in mammography without a palpable mass on physical examination[5] same as in our case. The patient's history of renal transplantation suggests chronic immunosuppression, which may have implications for disease development or progression. While a direct link between immunosuppression and MPD is not well-established, this case invites further exploration into potential associations, especially in immunocompromised populations. Our diagnosis was strengthened when biopsy results show cells with nuclear atypia and pale cytoplasm dispersed throughout the epidermis which are termed as Paget cells. Immunohistochemistry is useful in differentiating MPD from other clinical entities that it can mimic such as Pagetoid Bowen's disease, pagetoid melanoma, and pagetoid reticulocytosis. Paget's cells will also show the presence of other glandular antigens like EMA, CEA, gross cystic disease fluid protein 15, and various mucins.[9] Our patient tested positive for both EMA and CEA. Markers such as Cytokeratin 7 (CK7), GATA3, and HER2 are commonly expressed in Paget cells and aid in establishing the diagnosis. However, it is noteworthy that rare cases of CK7-negative MPD have been reported, highlighting the need for a panel of markers to avoid diagnostic pitfalls. The standard of care for MPD is mastectomy with or without axillary lymph node dissection.[10] The patient tested ER negative, PR negative, and HER2 positive. She underwent chemotherapy for 12 sessions followed by targeted therapy (Trastuzumab) for only 4 sessions due to intolerance of side effects. Initiation of appropriate treatment in our patient has prevented a possible dilemma such as metastasis to the lymph nodes. A more widespread disease will require more thorough tissue dissection and physical changes in the breast which may cause significant psychological distress. Regular follow-up appointments are essential to monitor for recurrence or development of new lesions. Patients treated with breast-conserving surgery followed by radiotherapy have a recurrence rate of about 8.0%.[1] Based on the NCCN Guidelines version 4.2025, the patient will be examined every 6–12 months for 5 years then annually. Management of MPD benefits from a team approach, involving a dermatologist, oncologist, surgeon, and radiologist to ensure comprehensive care. Patients with nonpalpable breast cancer have excellent survival outcomes, with studies reporting 5-year survival rates of 100% and 10-year disease-free survival rates between 92.6% and 94%, particularly among those with negative axillary lymph nodes.

MPD is not a preventable disease. It is crucial to recognize Paget's disease through clinical and pathological means as the superficial lesion might be the sole indication of an underlying ductal carcinoma. Moreover, its presence could carry important implications for the prognosis. Therefore, Paget's disease should be considered in cases of chronic eczematous breast lesions that are resistant to treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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