

# Paget's disease of male breast: A case report

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**Abstract** Paget's disease is an eczematous skin change of the nipple, usually associated with an underlying breast malignancy. Paget's disease of the breast is a rare entity with an incidence of 1.4-3% of all the mammary malignancies. Of all the malignant breast cancers, 1% occur in male patients, and thus Paget's disease of male breast is extremely rare. The most frequent presenting signs include ulceration, eczema, nipple discharge, bleeding, and crust formation. A case of Paget's disease of male nipple is reported in view of clinical rarity of this condition.

**Key words**

Paget's disease, malignancy, male breast.

## Introduction

Paget's disease is an uncommon and relatively rare intraepithelial adenocarcinoma presenting clinically as ulcerated or excoriated plaques.<sup>1</sup> Mammary Paget's disease (MPD) was discovered by James Paget in 1874 and extramammary type was found 15 years later by Radcliffe Crocker.<sup>2</sup> Mammary Paget's disease manifests as an underlying breast carcinoma that occurs in less than 5% of women suffering from breast cancer.<sup>3</sup> Some instances of Paget's disease of male breast have been cited in the literature.<sup>4</sup> Clinically patient presents with an eczematous skin lesion or persistent dermatitis of the nipple and the adjacent area often associated with burning sensation, ulceration, bleeding and oozing with serosanguinous discharge. Cutaneous examination reveals a scaly erythematous crusted plaque on the nipple and areolar skin. Retraction of the nipple or presence of palpable nodules indicates an underlying breast carcinoma. In most of the

cases, involvement is unilateral, however, bilateral mammary PD has been reported.<sup>5</sup> Regardless of the presence or absence of a palpable breast mass, modified radical mastectomy should be advised in all patients of biopsy-proven mammary Paget's disease.

## Case report

A 38 year old male presented with a 2 year history of itchy, reddish brown lesion on his right breast which was associated with burning sensation. The lesion had started from nipple and then progressively involved his entire areola. There was no history of nipple discharge, breast swelling or swelling in the axillary region. The patient was treated with multiple over-the-counter antibiotic-steroid creams prescribed by general practitioners with no improvement, rather steady progression to the present state. Cutaneous examination revealed a well-demarcated erythematous to brown colored scaly plaque measuring about 3×4cm over left breast involving the nipple and entire areolar area (**Figure 1**). There was no obvious nipple inversion, ulceration or active nipple discharge. There was no underlying palpable breast mass or any clinically palpable axillary or supraclavicular lymphadenopathy. The right

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Figure 1

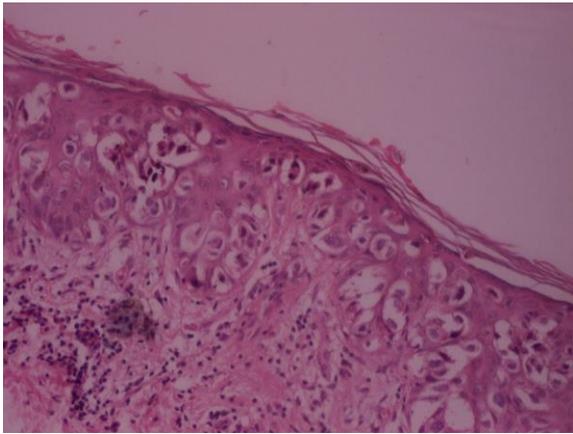


Figure 2

breast was normal both on inspection as well as on palpation. Mammography of the breast was done which did not reveal any parenchymal abnormality. A biopsy specimen taken for histopathological analysis revealed large atypical round to oval cells infiltrating the lower part of epidermis (**Figure 2**). The cells had a pale cytoplasm with prominent hyper chromatic nuclei, mitotic figures and were devoid of intercellular bridges. Immunohistochemistry was not done because of the lack of the facility. In view of clinical and histopathological evidence, a diagnosis of Paget's disease of male breast was entertained in this patient.

## Discussion

Paget's disease is an uncommon form of epithelial adenocarcinoma that involves mammary and extramammary tissues. Mammary Paget's disease is a rare disorder of the nipple-areola complex that is often associated with underlying in situ or invasive breast carcinoma.<sup>6,7</sup> The most frequent presenting signs include ulceration, eczema, nipple discharge, bleeding and crust formation.<sup>8</sup> Eczematoid changes of the nipple-areola complex and persisting soreness or itching, without obvious reason, is a suspicious symptom for Paget's disease of the breast and calls for a thorough evaluation, including mammography and surgical biopsy.<sup>6</sup> Paget's disease may present concomitantly with an underlying invasive carcinoma, ductal carcinoma in situ or with no underlying breast cancer. Contact dermatitis, Bowen's disease, Pagetoid dyskeratosis and malignant melanoma clinically may resemble Paget's disease but each of them have distinctive features on histology with the absence of Paget's cells. The histogenesis of mammary PD continues to be a matter of debate. The most accepted theory regarding the origin of Paget's cells is by migration from an underlying ductal carcinoma. An in-situ transformation theory has been proposed according to which the malignant cells can arise within the epidermis of the nipple independent of any pathological process within the parenchyma.<sup>9</sup> Poor prognostic factors include patients aging less than 60 years of age at the time of diagnosis (stage II) involved lymph nodes, underlying infiltrative carcinoma, palpable mass or multifocal lesions have lower prognosis. Histology may reveal hyperkeratosis, parakeratosis or acanthosis of the epidermis and infiltration with the classical Paget cell that is large, ovoid, has pale staining cytoplasm and hyperchromic nuclei.<sup>10</sup> Treatment is usually a mastectomy plus axillary node sampling or clearance. Adjuvant treatment may be

considered depending on nodal and receptor status.<sup>11</sup> Breast conservation surgery with radiotherapy, or radiotherapy alone, are not usually considered due to high recurrence rates.

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