Paget disease of the male nipple

ABSTRACT
Breast cancer occurring in the mammary gland of men is infrequent. It accounts for 0.8% of all breast cancers, which is less than one per cent of all newly diagnosed male cancers and 0.2% of male cancer deaths. However, Paget disease of the male nipple is extremely rare. We report a single case of Paget disease with infiltrative ductal carcinoma of the breast in a 61-year-old man.

KEY WORDS: Breast carcinoma, male nipple, Paget disease

INTRODUCTION
Since the initial description in 1874,[1] mammary Paget disease has remained a rare disease that accounts for 1 to 5% of all breast cancers and may be associated with underlying breast malignant neoplasm.[2] It is uncommon in men.[3] We would like to report a single case of Paget disease of the male breast, which we observed in our hospital.

CASE REPORT
A 61-year-old male was admitted for evaluation of a lesion on the left nipple. His nipple had become indurated, pruritic and with erythematous skin changes for the past year-and-a-half. Medical, family and social history records were unremarkable. Physical examination revealed a 3 cm, scaly, indurated plaque obscuring the normal architecture of the left nipple [Figure 1]. Lymphadenopathy was absent. He had no family history of breast cancer.

A mammogram showed a lesion highly suspicious of breast carcinoma [Figure 2]. The histopathologic diagnosis of biopsy of the lesion was infiltrative ductal carcinoma and Paget disease of the breast [Figure 3].

The patient underwent a left total mastectomy combined with axillary clearance of the nipple. The histology showed an invasive ductal carcinoma associated with Paget disease. The axillary nodes were free from metastases. Postoperative adjuvant therapy included chemo-radiotherapy. After two years, the patient continues to be disease-free.

DISCUSSION
Paget disease of the breast was first described, in 1874, by Sir James Paget in 15 women who presented with eruptions of the areola and nipple associated with subsequent breast cancer.[1]

Breast cancer occurring in the mammary gland of men is infrequent, accounting for 0.8% of all breast cancers; less than 1% of all newly diagnosed male cancers and 0.2% of male cancer deaths.[4]
The mean age of men who present with Paget disease of the breast is 60 years, but ages range from 43 to 81 years. The average delay in treatment of the disease from the onset of symptoms is about eight months in men. Although men present earlier with symptoms and show no histologic differences from women, their prognosis tends to be worse with a five-year survival rate of 20 to 30%.

Since the histological appearance of the Paget disease of the breast was first described, its histogenesis has been surrounded by much controversy. Two main hypotheses have tried to explain its nature and origin:

The first theory, the epidermotropic (ductal) theory, postulates that the Paget cells are, in origin, duct cancer cells that have migrated along the basal membrane of underlying ducts to the epidermis of the nipple. This theory is supported by the presence of an underlying intraductal or invasive carcinoma in majority of the patients.

The second theory, of in situ malignant transformation or degeneration from existing cells, regards the Paget cells as malignant keratinocytes appearing in situ and thus considers Paget disease of the breast to be an in situ carcinoma independent of any underlying carcinoma of the breast. This theory is supported by histologic studies in cases with the absence of an underlying carcinoma of the breast. Each of these theories is plausible; however, treatment approaches differ markedly depending on the theory of histogenesis.

Paget disease of the breast clinically presents as nipple erythema and mild eczematous scaling and flaking, progressing to nipple crusting, skin erosions and ulceration. The condition spreads outward off the nipple and onto the areola and surrounding skin of the breast. Temporary resolution of the eczematoid changes with or without the application of topical corticosteroids is possible and this may cause further delay in diagnosis. Paget disease of the breast may be asymptomatic and unsuspected clinically and may be reported as a histologic finding by the pathologist on a mastectomy specimen. Approximately 50% of patients also present with an associated palpable mass in the breast.

Paget disease is often mistaken for dermatitis or a benign dermatological condition involving the nipple. It is not unusual for the diagnosis to be delayed. A skin specimen containing Paget cells and a lactiferous duct secures the diagnosis and can be obtained by nipple scrape cytology or biopsy. More than 97% of patients with Paget disease have an underlying breast carcinoma. Paget may present with (54%) or without (46%) a mass. Invasive breast cancer coexists with Paget disease in 93% of patients with a mass and in 38% of patients without a mass. Because of the relative scarcity of cases in men, no studies have objectively compared treatment techniques in this population. Treatment may include modified radical mastectomy, axillary staging, radiation therapy, or systemic chemotherapies.

REFERENCES