Giant Paget's Disease of the Breast

– A case Report –

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We report a case of huge-sized Paget's disease of the breast with underlying invasive ductal carcinoma which showed no evidence of axillary lymph nodes metastasis.

A 73-year-old female patient noticed a small erosive eczematoid patch on the right nipple area 15 years ago. The lesion progressively enlarged, and at the time of visit, extended to form a 14 × 10 cm sized, well-demarcated, erythematous erosive patch with serosanguinous discharge and granulation tissue formation. The right nipple disappeared concomitantly. A firm, movable, 2.5 × 2.5 cm sized mass was palpated under her right breast.

Histopathologic findings of the biopsy specimen obtained from the skin lesion showed the permeation of the epidermis with numerous Paget cells. The dermis showed chronic inflammatory cell infiltration. In immunohistochemical staining, Paget cells were positive for carcinoembryonic antigen.

She was treated with a modified radical mastectomy. (Ann Dermatol 7(2):197–199, 1995)

Key Words: Breast, Giant Paget's disease

Paget's disease of the breast is a rare clinicopathologic entity characterized by progressive, marginated, eczematoid changes in the nipple and areola with abnormal cells within the epidermis. Lesion size ranges from 0.3 to 15 cm, with a mean 2.8 cm in one large series.

Paget's disease of the breast can be associated with an underlying carcinoma, usually intraductal in origin. This tumor may be small and noninvasive or, on the other hand, may be extensive and invasive with axillary lymph nodes metastasis.

We describe a case of Paget's disease of the breast showing a huge skin lesion with loss of the nipple associated with intraductal carcinoma of the breast.

REPORT OF A CASE

A 73-year-old woman had an extensive eczematoid lesion on the right breast. She had first noticed a small erosive eczematoid patch on the right nipple area 15 years ago. She had treated with topical agent intermittently, but the lesion slowly enlarged and extended to form a 14 × 10 cm sized, erythematous erosive patch with serosanguinous discharge and granulation tissue formation (Fig. 1).

The right nipple disappeared concomitantly. Her familial and past medical history were non-contributory.

Physical examination revealed a firm, movable, 2.5 × 2.5 cm-sized palpable mass on the right breast. A firm, non-tender, movable 2 × 2 cm-sized right axillary lymph node was also palpable. Laboratory evaluation revealed the results of VDRL, liver function test, urine analysis, and stool examination were all negative or within normal limits. Her leukocyte count was slightly elevated (12,500/μl). A chest X-ray film showed highly possible interstitial lung disease. Chest CT revealed mediastinal and right axillary lymph node enlargement with thickening of the right breast skin. Mammography suggested right breast cancer with diffuse breast skin thickening.

Histopathologic findings of a skin biopsy
Fig. 1. 14 × 10cm sized, well-demarcated, erythematous erosive patch with serosanguinous discharge and granulation tissue formation on the entire right breast. Loss of nipple is also seen.

Fig. 2. The epidermis shows large and round Paget cells which have pale-staining cytoplasm(H & E stain, × 100). Predominantly affected with an average age of approximately 55 years, although the disease may rarely be seen in men. The prognosis is worse in men. Although almost all cases of mammary Paget's disease are associated with underlying intraductal carcinoma of the breast, only 1.5-4.3% of the patients with breast cancer have Paget's disease.

Paget's disease of the breast occurs almost always unilaterally on the nipple and areola. Initial clinical changes may be manifested as erythema, scaling, or thickening of the nipple with later erosion and crust formation. The areola and surrounding skin may become involved as the well-demarcated surface lesion enlarges. Discharge, bleeding, ulceration, and occasional nipple invagination may also occur with pain, itching, or a burning sensation. Therefore if the patient has a unilateral eczematoid lesion on the nipple and areola recalcitrant to simple treatment, a thorough biopsy should be done to rule out the mammary Paget's disease. In the review of 109 female patients by Ascensao et al, lesion size ranged from 0.3 to 15 cm (in only 2 patients), with a mean 2.8 cm. Underlying breast mass can be palpated in over 28% of the patients.

Our case had a huge eczematoid lesion of 14 × 10 cm size, involving the entire surface of the right breast with the loss of the nipple. The loss of the nipple is thought to have been produced by the long-standing severe inflammation.

The characteristic histopathologic finding of mammary Paget's disease is the presence of Paget cells scattered throughout the epidermis in small numbers or in groups. Paget cells are large, rounded cells devoid of intercellular bridges, with ample, pale-staining cytoplasm. Occasionally, specimens showed the permeation of the epidermis with numerous Paget cells (Fig. 2). The underlying dermis showed chronic inflammatory cell infiltration. In immunohistochemical staining, Paget cells were positive for carcinoembryonic antigen (Fig. 3).

Histopathologic findings of the breast tissue and axillary lymph nodes revealed invasive ductal carcinoma with benign reactive lymph nodes changes.

She was treated with a modified radical mastectomy.

DISCUSSION

Paget's disease of the breast was initially described in 1874 by Sir James Paget. Women are
Paget cells contain some melanin, but they are do-pa-negative\(^1\). In immunohistochemical staining, carci-noembryonic antigen is regularly found in Paget cells\(^3\). Paget cells do not invade the dermis, which shows a moderately severe chronic inflammatory reaction\(^1\). Paget's disease of the breast should be differentiated from Bowen's disease and the superficial spreading type of malignant melanoma in situ. In both Paget's disease and Bowen's disease vacuolated cells may be observed, but clear -cut transitions between the vacuolated cells and epidermal cells, clumping of nuclei within multinucleated epidermal cells, and individual cell keratinization are observed only in Bowen's disease\(^4\). In addition, the cells in Bowen's disease do not contain carcinoembryonic antigen\(^1\). Superficially spreading malignant melanoma has pagetoid feature but may be differentiated by a positive reaction for S-100 protein or HMB-45 and negative reaction for carcinoembryonic antigen in immunohistochemical staining\(^1\).

The histopathologic findings of our case showed numerous Paget cells confined to the epidermis and these were positive for carcinoembryonic antigen in immunohistochemical staining.

The recommended treatments are modified radical mastectomy in those with a mass and a simple mastectomy for lesions with nipple involvement and no mass\(^6\). The important prognostic factor is axillary lymph node metastasis associated with clinically palpable breast mass. Ten-year survival rates were 9.9% for patients with positive nodes, whereas rates rose to 43.5% for patients with negative nodes\(^8\). Ten-year survival rates for patients with a palpable breast mass range from 22 to 33% in contrast to 82 to 91% for patients without a palpable breast mass\(^1\).

Our case had a palpable breast mass and was treated with a modified radical mastectomy. On histologic examination, the breast tissue obtained by mastectomy revealed the presence of intraductal carcinoma, and axillary lymph nodes showed benign reactive hyperplasia.

In summary, we present a case of Paget's disease of the breast associated with intraductal carcinoma of the breast, showing a huge eczematoid and erosive skin lesion with the loss of the nipple.

**REFERENCES**