

Paget's Disease of the Male Breast

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Paget's disease of the breast occurs almost exclusively in women, and is characterized by a unilateral, sharply defined eczematous skin lesion caused by epidermal metastases from underlying ductal adenocarcinoma of the breast. Paget's disease of the male breast is extremely rare.

A 61-year-old patient had a skin lesion on the right nipple for 1 year. The lesion was a well defined, brownish, crusted, and keratotic papule, and was associated with an underlying walnut sized, fixed, hard mass and a palpable ipsilateral axillary lymph node. Histologic findings were consistent with Paget's disease. Results of immunoperoxidase staining techniques for S-100 protein and carcinoembryonic antigen were negative. After a modified radical mastectomy and chemotherapy, the patient has been in remission for 12 months.

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Paget's disease of the breast represents an eczematous appearing process of the nipple and areola with cells containing large round nuclei and abundant pale-staining cytoplasm within the epidermis histopathologically. It occurs almost entirely in women, and is extremely rare in men.^{1,2} Elbogen in 1908 cited a case of mammary Paget's disease of male breast, and thereafter only 28 clinically and histologically confirmed cases have been reported.² In Korea, we could not find any cases in the literature.

We report a male patient with Paget's disease of the breast which has been confirmed histopathologically.

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REPORT OF A CASE

A 61-year-old man visited our department in April, 1990 with a pruritic eczematous skin lesion of the right nipple. For one year, he has been suffering from a recurrent eczematous skin lesion on the right nipple which has been treated with application of topical antibiotics. On initial physical examination, the patient seemed to be in good general health and had no recent history of weight loss. Examination of the right breast showed a well defined, pea sized, brownish, crusted, and keratotic papule (Fig. 1). On palpation of right breast, an underlying walnut sized, hard, fixed mass was noted. An enlarged, firm, movable lymph node was palpable in the right axilla.

The results of the following laboratory tests were within normal limits or negative: complete blood cell count, urinalysis, liver function test,



Fig. 1. A sharply defined, pea-sized, brownish crusted and keratotic papule and partially normal nipple of the right breast

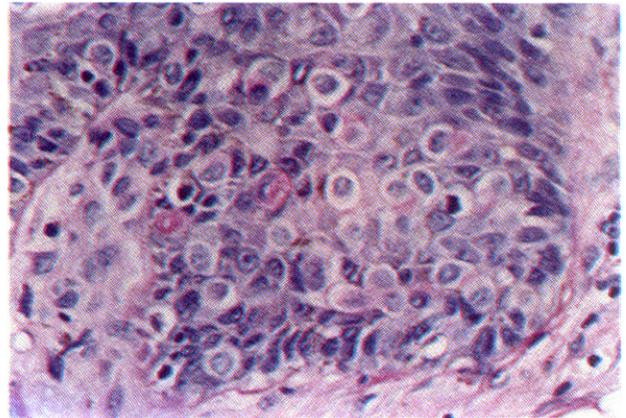


Fig. 3. Paget's cells stained red by the diastase-treated PAS stain (PAS stain, ×400)

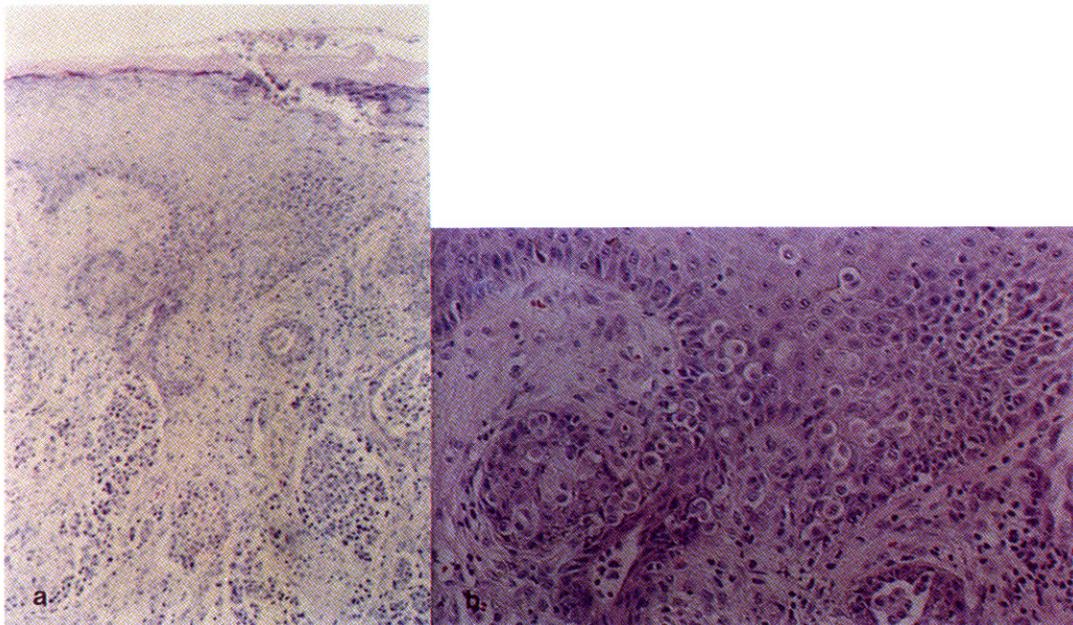


Fig. 2. Single or grouped, scattered, large, round Paget's cells with large, round nuclei and pale-staining cytoplasm in the epidermis, and ductal carcinoma in the dermis(H & E stain, (a) ×100, (b) ×200)

chest X-ray, electrocardiogram;no other evidences of distant metastases were found on the liver scan or bone scan. Histopathologic findings of the skin lesion revealed single or grouped pagetoid cells with large, round nuclei and pale-staining cytoplasm in the epidermis,

and ductal carcinoma in the dermis(Fig. 2). With special staining, diastase-treated PAS and alcian blue, a positive reaction occurred(Fig. 3, 4);while Fontana-Masson stain revealed negative findings(Fig. 5). Results of immunoperoxidase techniques by the Avidin-Biotin-Peroxidase

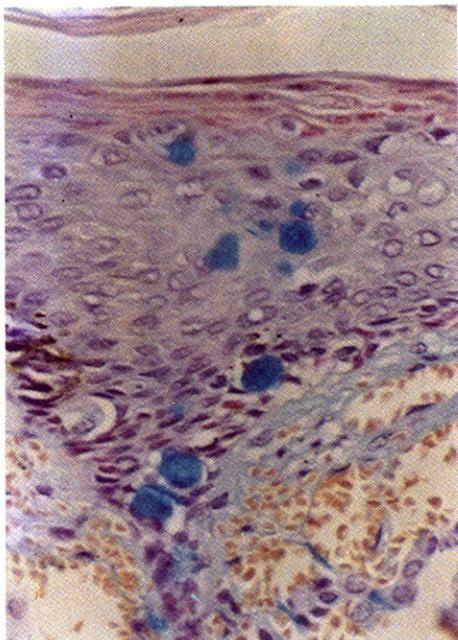


Fig. 4. Paget's cells stained blue by the alcian blue stain at pH 2.5(alcian blue stain, $\times 400$)

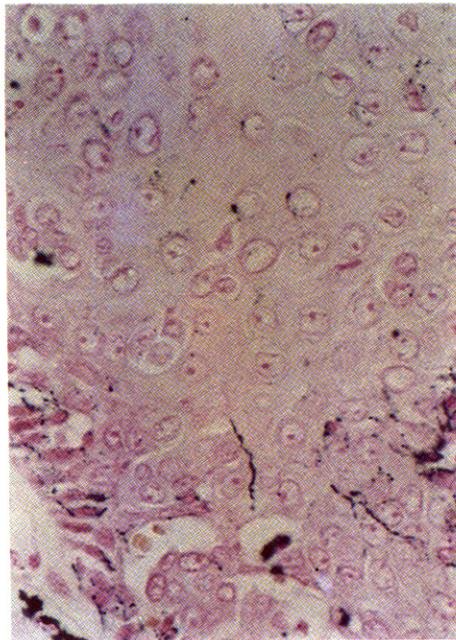


Fig. 5. Paget's cells unstained by the Fontana-Masson stain(Fontana-Masson stain, $\times 400$)

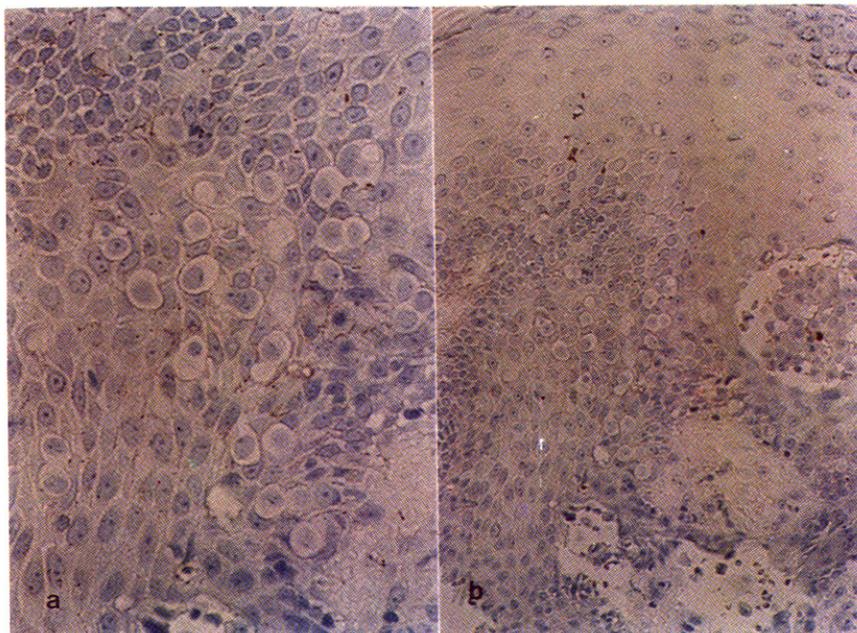


Fig. 6. a; Negative immunoperoxidase staining for S-100 protein($\times 400$),
b; Negative immunoperoxidase staining for CEA($\times 200$)

Complex method for S-100 protein and carcinoembryonic antigen were negative in the tumor cells (Fig. 6). Thus a diagnosis of mammary Paget's disease was made and the patient was transferred to the department of general surgery. A modified radical mastectomy was performed and the patient had 5 cycles of chemotherapy with cyclophosphamide, adriamycin and 5-FU. Sections of the underlying mass of the breast and right axillary lymph node showed glandular shaped atypical cells. The postoperative course was very good and the patient was followed for 12 months during which time he remained disease free.

DISCUSSION

Paget's disease of the male breast is an extremely rare clinicopathological entity. This is obvious from the low incidence of this disease, which comprises only 3 to 5% of all mammary malignancies, with an occurrence of less than 1% of all malignant breast tumors in males.^{3,4} Although mere 48 cases of Paget's disease of the male breast have been reported by various workers in the literature, only about half of them are documented with histopathological data.⁴

Collective review of relevant clinicopathological data on the reported 28 cases is as follows^{2,3}: The most frequent site was the nipple; the most frequent clinical signs and symptoms were ulcerations and eczematous changes; next were lumps of the breast with discharge and bleeding, itching or pain, and induration. A lump of the breast was found in 12 cases (42.8%). Palpable enlarged axillary lymph nodes were present in 15 cases (53.5%). Nipple erosion with a palpable lump of the breast is typical in the majority of the cases; the former may precede the development of a detectable mass by many years. The age of the patient varied from 43 to 81 years, with a predilection between 50 and 70 years of age.

The present case presented with a unilateral, recurrent eczematous skin lesion with discharge and bleeding. The patient had done well without specific treatment, but was admitted for consultation during which time the palpable mass of the breast was found. Hutchin and Houlihan⁵ classified Paget's disease of the male breast into three clinical types: (1) Paget's disease associated with nipple erosion only, (2) Paget's disease associated with nipple erosion and a palpable breast tumor, and (3) Paget's disease associated with a breast tumor alone. The present case belongs to the second clinical type.

Histopathologic findings were diagnostic for mammary Paget's disease cells which have a large nuclei and abundant pale-staining cytoplasm. Mammary Paget's disease must be differentiated from Bowen's disease and the pagetoid type of malignant melanoma in situ.^{1,6-9} Paget's cells contain mucin that stains with diastase-treated PAS and alcian blue at pH 2.5 with a positive staining reactions. Occasionally, Paget's cells contain some melanin, but these cells do not stain with dopa. S-100 protein may identify benign and malignant melanocytic lesions, Langerhans cells, and cells of Schwann cell origin. Anticarcinoembryonic antigen antibody identifies mammary and extramammary Paget's cells and cells of apocrine and eccrine origin. In general, mammary Paget's disease has a negative reaction for S-100 protein and a positive reaction for carcinoembryonic antigen, but there are exceptions.¹⁰⁻¹² The present case showed a negative staining reaction with the Fontana-Masson stain, but was positive with diastase-treated PAS staining and alcian blue staining at pH 2.5. Results of immunoperoxidase staining techniques for S-100 protein and carcinoembryonic antigen were negative. Thus, we could differentiate it from Bowen's disease and the pagetoid type of malignant melanoma in situ.^{8,13}

Treatment of choice for mammary Paget's

disease is a surgical excision. Adjunctive therapies include postoperative radiation, chemotherapy, orchidectomy, adrenalectomy, hypophysectomy, tamoxifen, and hormonal therapy.^{2,8} In the present case, a modified radical mastectomy and combined chemotherapy were done, and the patient has done well up to the present time.

Prognosis of mammary Paget's disease in men is poorer than that of women, but both appear to have a poor prognosis, particularly if a lump is present. The estimated 5-year survival rate for female patients is approximately 45%, and in the male patients, 20% to 30%.^{2,10} These results may be due to the following factors: (1) the hormonal environments are quite different in each sex, (2) medical attention is usually delayed in the male patient, so that the disease is quite often far advanced when it is first diagnosed, and (3) because the male breast is smaller than the female, the tumor tends toward early infiltration of the underlying structures, skin, and regional lymph nodes.^{6,13}

Dermatologist should do biopsy on every eczematous skin lesions of the nipple and areola. The earlier this procedure is done, the more favorable the results are.

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