

Sema HÜCÜMENOĞLU¹
Handan KAYA²
Fatih Mehmet AVŞAR³

Paget's Disease of the Male Breast - a Case Report

¹Department of Pathology, SSK Ankara Training and Research Hospital, Ankara.

²Department of Pathology, Faculty of Medicine, Marmara University, Istanbul.

³Department of 1st Surgery, Ankara Numune Training and Research Hospital, Ankara - TURKEY

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Paget's disease of the breast is a rare condition with an incidence of 1-4.3% of all mammary malignancies. Of all malignant breast cancers, 1% occur in male patients, and thus Paget's disease of the male breast is extremely rare (1,2,3).

In this report, we study a fully documented case of Paget's disease of the male breast.

Case report

A 74-year-old man presented with a lump in the left breast of 6 months duration and ulceration of the nipple. No family history of breast cancer was present.

On physical examination, the nipple of the left breast was retracted and ulcerated. A 6x6 cm² hard, irregular and nontender subareolar mass was palpable. A few firm mobile lymph nodes with 0.5-1 cm enlargement were palpable in the left axilla. The patient underwent to tru-cut biopsy. The histologic examination of this biopsy showed an infiltrating carcinoma. After that a modified radical mastectomy was performed.

Macroscopically, there was a firm gray-white mass with 6x6 cm enlargement just below the nipple area. Microscopic examination showed disorganization of the cellular arrangement of the epidermis with accumulation of large atypical epithelial cells showing clear cytoplasm and atypical nuclei (Figure 1). These cells were scattered singly or in clusters among normal looking cells

continuous with the tumor beneath (Figure 2). The axillary lymph nodes showed no tumor metastases. The tumor that was seen macroscopically beneath the areola was microscopically an infiltrating ductal carcinoma. Atypical cells within the epidermis stained moderately positively with acidic mucin but negatively for periodic acid-Schiff (PAS). Immunohistochemistry was performed by the immunoperoxidase technique on the paraffin embedded tumor tissue. Estrogen receptor expression was negative by immunohistochemistry. Epithelial membrane antigen (EMA) and low molecular weight cytokeratin showed strong immunoreactivity in abnormal cell population within the epidermis (Figure 3,4). While tumor cells exhibited weakly positive immunoreactivity with S-100 and carcinoembryonic antigen (CEA), they were entirely negative with HMB-45. Therefore a diagnosis of Paget's disease of the nipple was made. It is now 14 months postoperative and the patient is alive without any evidence of recurrence.

The rarity of Paget's disease of the male is obvious in a survey of the literature. Until 1969, Crichlow and Czernobilsky accepted only 11 cases as genuine Paget's disease and reported their two own cases (1,4). Coley and Kuehn reviewed the literature up to 1971 and published five cases of their own (1,5). Serour reviewed the data of 28 cases including theirs (1). The diagnosis of Paget's disease is established by histology. Typically it is associated with an underlying intraductal or infiltrating

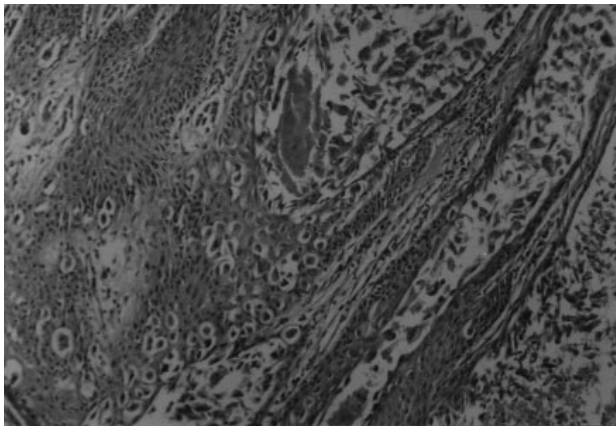


Figure 1. Paget's cells scattered in the epidermis, also seen in continuity with the ductal carcinoma (H&E x 100).

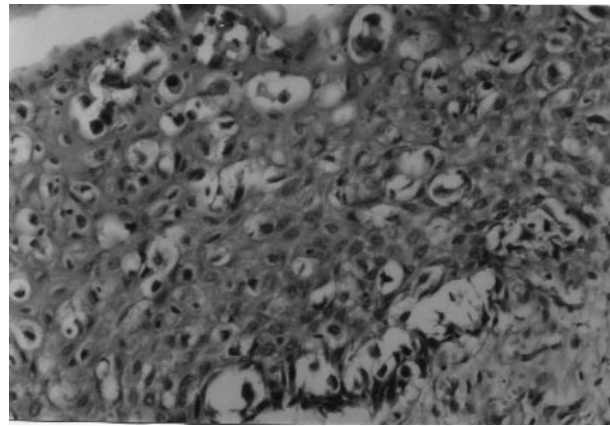


Figure 2. Higher magnification showing Paget cells in small clusters within the epidermis (H&E x 400).

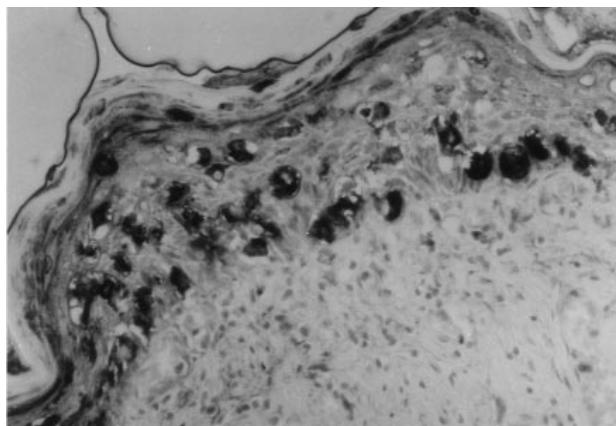


Figure 3. Immunoperoxidase stain for low molecular weight cytokeratin strongly stained abnormal cell population (H&E x 200).

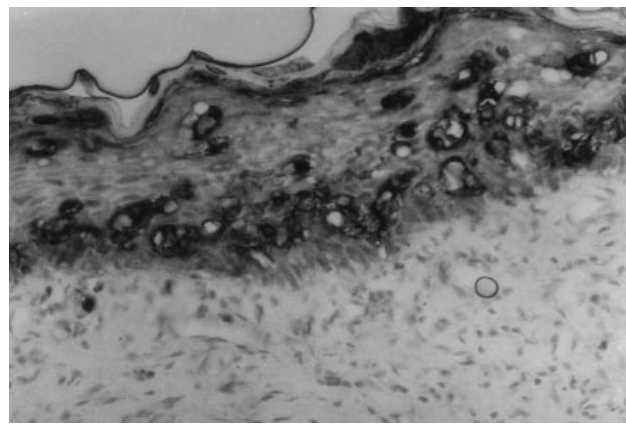


Figure 4. Immunoperoxidase stain for epithelium membrane antigen strongly stained Paget cell (H&E x 200).

ductal carcinoma. O'Sullivan (6) and Takeuchi (7) reported two histologically confirmed cases of Paget's disease of the breast without an underlying carcinoma. When entertaining the diagnosis of Paget's disease of the breast using morphological characteristics of the cells, the diagnosis of malignant melanoma, Bowen's disease (intraepithelial squamous cell carcinoma), superficial spreading basal cell carcinoma, dermatitis, clear cell papulosis and Toker's cells must be included in the differential diagnosis (1,3,8,9). Histochemical stains have been found to be very useful for the diagnosis of Paget's disease (1,2). The accurate diagnosis of Paget's disease is possible by immunohistochemistry.

Paget's disease usually presents in the 5th and 6th decades of life (3,6,8,10). The average delay between the

onset of signs or symptoms and the institution of definite treatment averages approximately 8 years, compared with 12 to 14 months in Paget's disease of the female breast (3,10).

The most frequent presenting signs include ulceration, eczema, nipple discharge, bleeding and acrust formation (1,3,8,10).

Two major histogenetic theories of Paget's disease have been proposed. The more accepted theory is the origin of the Paget cell from the ductal carcinoma spreading to the nipple by either metastasis or upward migration (3,9,10,11). The second theory holds that the Paget cell is an epidermoid cell transformed in situ without a direct connection with the underlying carcinoma (3,9,10,12).

Although there seems to be no significant histological difference between tumors in male and female patients, the former appears to have a poorer prognosis, particularly if a lump is present (1,10,13). Carcinoma with Paget's disease carries a poorer prognosis than ordinary male breast cancer (10). The estimated 5-year

survival rate for male patients is approximately 20% to 30% (1,3,10).

In conclusion, we report a classic case of Paget's disease of the male breast with underlying infiltrating ductal carcinoma and review the literature.

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