Paget's disease of the Male Breast with Underlying Ductal Carcinoma in Situ

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KEY WORDS: Paget's disease, breast cancer, male breast cancer, ductal carcinoma in situ (DCIS)

IMAJ 2015; 17: 64–65

Male breast cancer is rare, accounting for approximately 1% of all breast cancers [1,2]. The common patient is in his sixth decade of life with a genetic predisposition to Klinefelter's syndrome, a positive family history of breast cancer, or *BRCA2* mutation [1]. Among male patients with mammary carcinoma, Paget's disease of the nipple compromises 1–3% of all breast cancers and its appearance among male patients in extremely uncommon [4]. We report a case of histologically confirmed Paget's disease of the breast in a 63 year old man and discuss the appropriate diagnostic and treatment strategies.

PATIENT DESCRIPTION

A 63 year old Caucasian male patient with a medical history of mental retardation and schizophrenia was referred to our surgical outpatient clinic with scaling and erythema of the right nipple areola complex (NAC). No information regarding the duration of the lesion or family history of breast cancer could be attained. Examination of the right breast revealed an ulcerated erythematous lesion with scaling confined to the NAC. On gross physical examination of the breast there were no signs of nipple retraction or discharge. A mobile mass measuring 2 cm in diameter at the sub-areolar plane was identified. There was no evidence of lymphadenopathy, and the left breast was normal. Breast ultrasound examination was normal.

Due to the differential diagnosis of skin and subcutaneous disorder, a punch biopsy from the lesion was performed. Histological examination of the 0.3 cm cylindrical specimen revealed Paget's disease of the breast, which was supported by positive staining with cytokeratin 7 (CK 7) and negative for melanoma antigen recognized by T cells 1 (MART1) as well as human melanoma black 45 (HMB 45). The patient was scheduled for a mastectomy and sentinel lymph node biopsy. In view of the patient's limited mental functioning, and acceding to the family's request, the operation was directed into local disease control with further procedures subject to the final pathologic report. The patient was admitted electively to our surgical department 1 month after the outpatient clinic visit. On physical examination the lesion had expanded radially to involve the peri-areolar skin [Figure 1] with a diameter of 4.5 cm. Physical examination and blood tests were unremarkable. Chest X-ray and electrocardiogram were within normal limits. After obtaining guardian consent, we performed a simple mastectomy. His postoperative course was uneventful and he was discharged after 3 days.

The mastectomy specimen was elliptical and measured 10.5 x 10 x 4 cm, with central ulceration at the NAC area, measuring 4.5 cm in diameter. In the sub-areolar plane a white and rigid focus of tissue measuring 2 cm in diameter was identified. Histological examination showed Paget cells at the NAC, with two foci of ductal carcinoma in situ, solid type. The surgical margins were free of tumor. Immunohistochemistry staining of the Paget cells was positive for cytokeratin MNF 116 (CK-MNF 116), cytokeratin 7 (CK 7) and cytokeratin 8/18 (CK 8/18). The Paget cells were also positive for estrogen receptors and carcinoembryonic antigen (CEA), and negative for cytokeratin 20 (CK 20), progesterone receptors, and human

Eczematoid lesion of the right nipple areola complex

melanoma black 45 (HMB 45). Ductal carcinoma in situ immunostaining was positive for estrogen receptor and negative for progesterone receptor. Her-2/neu immunohistochemical examination showed faint weak partial membrane staining in 80% of the tumor cells. The patient was referred to the oncologist for further treatment.

COMMENT

The patient was referred to our department with an eczamatoid lesion involving the NAC. Differential diagnosis includes, by descending order, eczema, Bowen's disease, squamous cell carcinoma, melanoma and Paget's disease. Histological evaluation confirmed the diagnosis of Paget's disease. Paget's disease does not penetrate beyond the basement membrane and is therefore defined as carcinoma in situ. However, in more than 90% of cases there is ductal carcinoma in situ (DCIS) or invasive breast cancer in the underlying breast tissue [3]. In our patient the sub-areolar mass was diagnosed as DCIS compared to other reports where palpable mass usually involves invasive breast cancer [3].

Treatment consists of mastectomy and sentinel lymph node biopsy, although several cases of lumpectomy accompanied by radiation and adjuvant therapy have been reported [1]. In males, surgical choices are the same as for females, as mentioned above. In our patient, in view of his personality disorders, treatment comprised mastectomy only, with optional staged lymph node biopsy. Prognostic factors are the same as for female breast cancer and include nodal involvement and palpable mass size. The 5 year survival rate of male patients with Paget's disease is 20–30% compared to 30–40% in females [4]. The presence of DCIS or invasive breast cancer alters the survival rate, as does palpable mass [5]. The average delay between treatment and diagnosis is 7–8 months [1], unlike the 3 months in our patient.

We wish to draw the attention of physicians caring for patients with nipple dermatitis, eczematoid or psoriatic lesions to this lethal and rare tumor.

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