



Pagets Disease Of The Nipple

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Case Discussion

A 38-year-old female diagnosed and treated for wet eczema of left nipple for 3 months was referred for further management due to failure of the dermatological management. The patient had no other symptoms and general examination was unremarkable. Local examination of the left breast revealed weeping dermatitis involving the nipple-areola complex and the surrounding skin circumferentially (Illustration 1). There were no palpable lumps in both breasts or lymph nodes in both the axilla. A clinical diagnosis of Pagets disease of the nipple was made and a skin pinch biopsy was performed which confirmed the diagnosis for which she underwent a modified radical mastectomy. Final histopathology reconfirmed the diagnosis with 3/14 positive lymph nodes (Illustration 2). She received adjuvant chemoradiation and at the end of 3 years of follow-up she is well with good locoregional and systemic control.

Pagets disease of the nipple presents as an eczematous lesion, occurs in 1–4% of all female breast carcinoma cases and is invariably associated with underlying malignancy either overt or occult. Majority of these patients have an invasive disease although 40–45% are associated with DCIS [1]. Paget's disease of the breast was first described by Sir James Paget in 1874 who described "an eczematous change in the skin of the nipple preceeding an underlying mammary cancer"[2]. It is a red, oozing, crusted lesion which is often unresponsive to topical steroid and antibiotics. Histopathologically, there is a proliferation of malignant epithelial cells scattered throughout the epidermis. The cells have abundant pale staining cytoplasm surrounding a hyperchromatic nucleus with prominent nucleoli [3]. Opinion is divided regarding the pathogenesis of this condition. It is thought that malignant epithelial cells from intraductal carcinoma extend into the overlying epidermis through mammary duct epithelium and proliferate in the epidermis causing thickening of the nipple and areolar skin. This is supported by the observation that Paget cells often share cell surface markers with the underlying breast carcinoma (e.g. CAM 5.2, CEA, c-erb 2 and EMA) [3, 4]. Normal epidermal keratinocytes produce and release the mobility factor heregulin-alpha which is chemotactic for heregulin receptors (Her-2) and coreceptors Her 3 and

Her 4 which are produced by Pagets cells. This is thought to result in migration of these cells to the nipple epidermis [5]. Others believe that Paget's cells are derived from clear nipple epithelium cells and that underlying intraductal carcinoma is simply coexisting with this disease [4].

Eczema is the major differential diagnosis. Other differential diagnoses include contact dermatitis, frictional hyperkeratosis, psoriasis, bacterial, viral or fungal infection. Malignant diseases such as Bowen's disease, superficial basal cell carcinoma, melanoma or skin metastasis should be excluded [1].

The treatment of breast Paget disease is related to the coexistence of the underlining breast carcinoma. In cases with palpable breast cancer, the only choice is considered to be radical mastectomy (either classic or modified) with axillary node excision. When there is no palpable tumor one may try conservative interventions, with or without node excision. Studies shows in those cases reoccurrences after a median period of 4.6 years [6]. The survival is correlated with presence/absence of the palpable breast tumor. The overall rate of survival is 38-40% at 5 years and 22-33% at 10 years in patients with associated breast tumor. The death rate in patients with metastatic breast carcinoma and Paget disease is 61.3%, with a cumulative rate at 10 years of 33%. The survival rate in patients with Paget disease and no associated breast malignancy is 92-94% at 5 years and 82-91% at 10 years [7].

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Illustrations

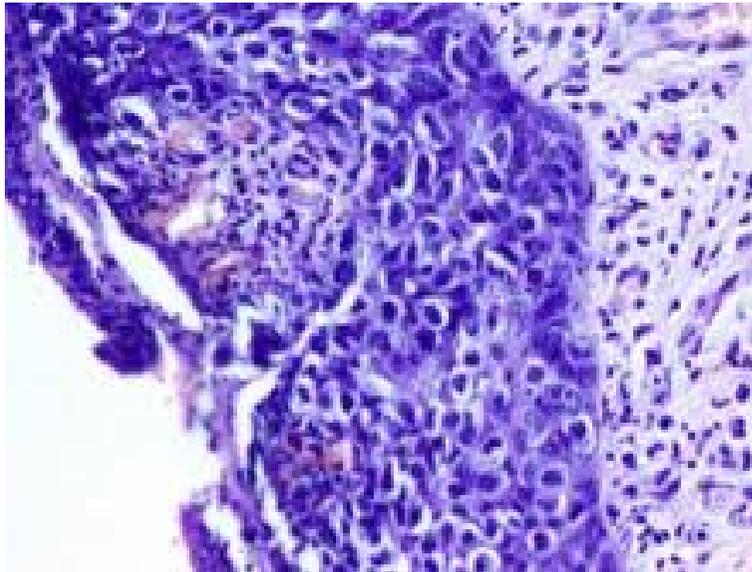
Illustration 1

Eczematous lesion



Illustration 2

Histopathology



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