

Bilateral Paget's Disease of Nipple

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Summary:

Paget's disease of nipple is a rare entity of malignant lesion of breast and bilateral Paget's disease is very uncommon. It presents as eczema like condition of the nipple areola which persists despite local treatment. As it is a superficial manifestation of underlying breast carcinoma high degree of suspicion is essential for proper recognition of this condition, so that early diagnostic work up can be initiated for differentiating it from other benign inflammatory skin

Introduction:

Paget's disease of nipple is uncommon type of breast carcinoma. More than 95% of patients also have underlying invasive or noninvasive breast carcinoma.¹ A palpable mass in breast may or may not be present. Usual presentation is as a chronic eczematous eruption of the nipple progressing to ulcerated, red weeping lesion. Paget's disease is frequently confused with eczema which tends to occur in younger people having signs of eczema elsewhere in the body. Eczema primarily involves the areola and only secondarily affects the nipple and mostly bilateral. Whereas Paget's disease is

disorder and for detecting an underlying breast carcinoma, thereby delay in proper treatment can be avoided in order to avoiding dismal consequences. Here we report a case of bilateral Paget's disease of nipple with invasive left breast carcinoma in a 40 year old lady diagnosed in Medical College for Women and Hospital, Uttara, Dhaka.

Key words: Paget's disease, Breast carcinoma.

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usually unilateral, always affects the nipple first then involves the areola as a secondary event.² Although rare, Paget's disease of nipple can occur in both breasts. In our case report the patient developed Paget's disease in one breast and treated as benign skin lesion for 3 years. She noticed a lump in same breast 3 & ½ years later and developed Paget's disease in other breast. This report focuses on the fact that though this condition is often innocuous and limited to a surface appearance, actually indicative of a very serious underlying breast carcinoma.³ So early diagnosis and prompt treatment is the basis for better outcome.

Case report:

A 40 year old lady, housewife with lower middle socio economic background presented to us on June 2010 with recurrent itching and excoriation of left nipple areola for 4 years. Lesion started at nipple then extended to areola. She experienced same at right nipple for 4 months. She also noticed a painless lump in left breast for 4 months which was gradually increasing in size. She had no nipple discharge and did not give any previous history of breast diseases. The patient had no history of eczema or dermatitis elsewhere in the body. She received treatment as eczema and fungal disease but did not improve completely. She was a premenopausal lady and her menarche started at her 13 years of age. She was a mother of 3 children; all were breast fed, her age was below 25 years during first baby;. She did not take any contraception. She was normotensive and nondiabetic. Her appetite was good and she had no

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recent weight loss. She had no history of cough, haemoptysis and jaundice or bone pain. She had no family history of breast cancer or any other cancer. She is neither a smoker nor an alcoholic. She used to take average Bengali foods.

Examination of her breasts revealed moderate excoriation of nipple with erythema in right breast without any palpable lump and severe excoriation of nipple areola in left breast with swollen areola. A well defined hard, nontender, irregular lump of about 3x3cm was found in upper and outer quadrant of left breast encroaching to areola with no fixity to underlying structures or overlying skin. Surrounding skin of her both breast appeared normal. A mobile hard, nontender lymph node of about 2x2cm was palpable in left axilla. Her right axilla and supraclavicular fossa were free of palpable lymph nodes.



Fig.-1: Right breast



Fig.-2: Left breast

She was of average weight and height with sound mental status and unremarkable general status. Her breath sound was normal; she had no hepatomegaly or ascites. No abnormality was found by digital rectal and per vaginal examinations.

FNAC of her left breast lump showed invasive duct cell carcinoma, FNAC of her left axillary lymph node revealed no malignant cell. Histopathological examination of incision biopsy from right nipple and left nipple areola confirmed Paget's disease. Ultrasound scan of both breasts detected a echogenic irregular mass in left breast. Bilateral mammography suggested a suspicious mass lesion in left breast with left axillary lymphadenopathy. Her chest skiagram was normal, ultrasound scan of whole abdomen showed cholelithiasis. Her CA 15-3 level was within normal limit. She had normal bone scan. Other routine investigations were unremarkable.

She underwent simple mastectomy with axillary dissection on left side and simple mastectomy on right side. Histopathology of tissue from left breast lump showed invasive ductal carcinoma(NOS) with invasion into surrounding tissue, grade – 3, Bloom- Richardson score 8/9. Resection margins were free of tumor, 3 Lymph nodes out of 8 showed metastatic adenocarcinoma with involvement of adjacent fibrofatty tissue. No underlying malignancy was found in excised right breast. Receptor analysis of specimen revealed negative for estrogen & progesterone receptor with positive for HER – 2.

Her post operative period was uneventful. Subsequently she delayed in receiving adjuvant therapy. After 4th cycle of chemotherapy her general condition was deteriorating; multiple secondary lesions in liver were evident on ultrasonogram. She survived for 9 months after operation.

Discussion:

Paget's disease of breasts represents approximately 1-3% of all breast malignancies; Sir James Paget, an English surgeon first described it in 1874.³ It is usually associated with Duct cell carcinoma in situ or invasive carcinoma. More than 50% of patient have lump in breast. It is most common in women in their 50s but can occur at a younger or older age. Involvement of male breast is rarely reported.⁴ Only a few cases of bilateral Paget's disease of the breast have been described world wide.⁵ To our best knowledge no such case was reported in

Bangladesh. In a study in Japan Paget's disease accounts for 1.2% of all breast cancer.⁶ In a case report, Bijian xie et al described bilateral paget's disease of the breast without underlying cancer in a 45 year old Chinese lady having gastric cancer.⁵

Exact cause of Paget's disease is still unknown. But in this connection two theories have been suggested in literature. One theory proposes that cancer cells called paget cells break off from a tumour inside the breast and move through the milk ducts to the surface of the nipple resulting in Paget's disease of nipple. The theory is supported by the fact that more than 97% of the patient with Paget's disease also have underlying breast carcinoma or duct cell carcinoma in situ. Other theory suggests that skin cells of nipple spontaneously become Paget cell. This theory is supported by the rare case of Paget disease in which there is no underlying breast carcinoma and the cases in which the underlying breast carcinoma is found to be a separate tumour from paget's disease.¹

Initially Paget's disease presents as mild eczematous rashes or scaling with redness & itching affecting usually only one nipple. Then the condition spreads outward onto areola & surrounding skin of the breast.⁷ More advanced disease may show more serious destruction of skin with straw coloured or bloody nipple discharge; nipple becomes flattened or inverted or is eroded slowly and eventually disappear leaving a weeping ulcer.⁸ Underlying breast carcinoma sooner or later becomes clinically evident. Patient may have pain, tingling, itching, burning and increase sensitivity. It is common for the symptoms to disappear for a while which misguides the patient.

The clinical differential diagnosis of this condition includes eczema, contact dermatitis, psoriasis and post radiation dermatitis.⁷

An incision biopsy from the lesion can confirm the diagnosis by the presence of paget's cell – large, pale, vacuolated cells in the epidermis. Paget's cells do not invade through dermal basement membrane therefore are a form of carcinoma in situ.⁷ Other useful diagnostic tools are imprint or scrap cytology or superficial epidermal shave biopsy or punch biopsy of nipple lesion. Sample of nipple discharge may also be examined microscopically for presence of paget's cell.⁹ Fine needle

aspiration cytology of the lump confirms the underlying breast cancer where lump is present. Mammography may be helpful for better delineation of any abnormalities like skin thickening, nipple retraction, presence of subareolar mass, subareolar or diffuse malignant calcification, a discrete or multifocal mass.¹⁰ Supplementary ultrasonogram of breasts is helpful for further evaluation of abnormal mammographic findings. Detection of early stage of cancer by Magnetic resonance imaging is also reported.¹¹

Surgical treatment is the mainstay of treatment. Simple mastectomy with or without an axillary node procedure is recommended for noninvasive carcinoma. Extensive duct cell carcinoma in situ or invasive carcinoma is treated by modified radical mastectomy. Anticancer drugs, radiation therapy, hormone therapy or immunotherapy are the other modalities of adjuvant treatment.

Survival rate in paget's disease alone at 5 year is 85% when lymph node is negative whereas in cases of positive lymph node it is about 32%. Patient with an underlying breast carcinoma have survival rate of 38 – 40% at 5 years. Death rate of metastatic breast carcinoma in patient with paget's disease & underlying carcinoma is about 61.3%.¹² Our patient got the dismal outcome probably due to delay in diagnosis and subsequently delay in starting chemotherapy.

Conclusions:

According to cancer research UK, paget's disease is found in one or two out of every 100 cases of breast carcinoma.¹³ There are fewer than 10 cases of bilateral involvement described in the literature.⁵ It is difficult to diagnose due to its resemblance to dermatitis & eczema. Histopathological study is the only gateway. Awareness of women is vital. Early diagnosis ensures better outcome. So the women experiencing any changes in nipple areola always should get examined by their General practitioner and who should not delay to refer the suspicious cases to specialist doctor.

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