RESEARCH PAPER | Medical Science | Volume : 4 | Issue : 4 | Apr 2014 | ISSN - 2249-555X

Paget's Disease of Breast : A Clinical Dilemma

KEYWORDS: Breast, Paget, mastectomy, carcinoma, lymphadenopathy

Dr. Richa Jaiman
Associate Professor, Department of Surgery, S.N. Medical College, Agra, U.P.

Dr. Puneet Srivastava
Assistant Professor, Department of Surgery, S.N. Medical College, Agra, U.P.

Dr. Parveen Kumar
Junior Resident-IIIrd Year, Department of Surgery, S.N. Medical College, Agra, U.P.

Dr. Kuldeep Singh Yadav
Junior Resident-IInd Year, Department of Surgery, S.N. Medical College, Agra, U.P.

ABSTRACT *Paget's disease is a rare breast malignancy. The clinical entity of paget's disease is distinct among breast carcinomas but is usually confused with benign conditions like contact dermatitis, eczema, drug reactions or psoriasis. The journey from differential diagnosis to provisional to definitive diagnosis is not easy and require multiple modalities and patience. Here we present a case of clinical interest which later turned to be paget disease with underlying invasive ductal carcinoma with metastatic lymph nodes and underwent modified radical mastectomy in our hospital.*

INTRODUCTION:
Paget's disease accounts for 1% or less of breast malignancies, characterized clinically by nipple erythema, crusting ,ulceration with extension into areola and surrounding skin. It was first described by Sir James Paget in 1874. Carcinoma cells invade across the junction of epidermal and dermal epithelial cells and enter epidermal layer of skin of nipple and surrounding tissue. More than 95% of patients with paget's disease have an underlying breast carcinoma. Paget's disease may have accompanied palpable mass in over 50%. Invasive breast cancer will be identified in patients with a palpable mass and paget's disease in over 90% of patients. This article highlights the clinical dilemma and approach to paget's disease.

CASE REPORT:
A 40 year female presented to surgery OPD with complain of some skin lesion over left breast since 1 year. There was no history of trauma, any discharge or any other swelling in breast. History of slight itching on and off was present. On clinical examination, an erythematous lesion involving nipple, areola and surrounding skin was noted with irregular margins, slightly retracted nipple and encrustations over the lesion. [Fig.1].

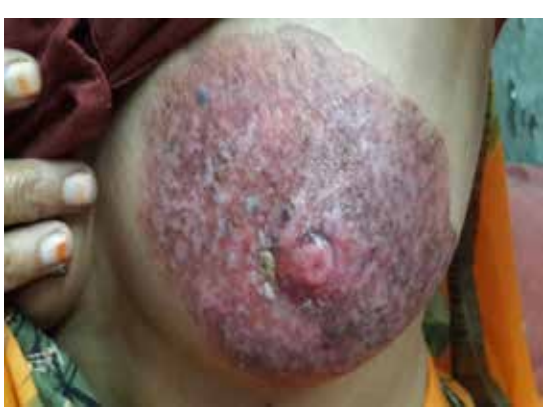

Fig.1. Paget's disease skin lesion.
There was no palpable abnormality in breast and no axillary lympadenopathy. The contralateral breast was normal. A dermatology opinion was taken and after clinical discussion, differential diagnosis of paget's disease, eczema, tinea corporis and psoriasis were made. The patient was prescribed a two week course of antibiotics and topical application. Mammography (MMG) showed inflammatory sequelae. [Fig.2,3].

Fig.2. Mammography findings on cranio-caudal view.

Fig.3. Mammography findings on medio-lateral view.

Volume : 4 | Issue : 4 | Apr 2014 | ISSN - 2249-555X

After two weeks, there was no change in lesion and a skin biopsy was advised which revealed stratified squamous epithelium with mild dysplastic changes in basal layer and mild to moderate lymphocytic infiltrate in sub epidermal region. A provisional diagnosis of paget's disease was made and patient underwent left modified radical mastectomy and the specimen was sent for histopathological examination and report was of paget's disease with underlying invasive ductal carcinoma and metastatic foci in lymph nodes. Patient was referred to radiotherapy department for chemotherapy.

DISCUSSION:

Paget's disease is a rare breast malignancy, usually with underlying ductal carcinoma in situ. It occurs most commonly in post-menapousal women, often during the sixth decade of life.[1] Pathologically a paget cell is a large, pale staining cell with round or oval nuclei and large nucleoli. It does not invade basement membrane and are categorized as carcinoma in situ. The clinical presentation of dermatitis like picture confuses the diagnosis. A history of lesion involving nipple first and then extending to areola and surrounding skin, gives a clue to paget's disease. This history was not clear cut in our patient and so a dermatological opinion was sought. History of any drug reaction was eliminated and a course of two weeks antibiotic was prescribed along with topical steroid application, to rule out dermatitis due to tinea corporis or psoriasis. There was no palpable abnormality in left breast of our patient. Skin biopsy was judgemental with dysplastic changes and patient was proceeded through clinical diagnosis of paget's disease. The histogenesis of paget's disease is debatable. The epidermotropic theory says that paget's cells are ductal carcinoma cells that have migrated from the underlying breast parenchyma to the nipple epidermis. According to the in situ transformation theory, the paget's cells arise as malignant cells in the nipple epidermis independent from any other pathologic process within the breast parenchyma. [2] This theory has been proposed to explain those cases in which there is no underlying mammary carcinoma or when there is a carcinoma remote from the nipple-areola complex.

MMG is not always a reliable procedure for detecting MPD (Mammary Paget's Disease). It is limited in its depiction of underlying DCIS in women with paget's disease. The sensitivity of MMG seems to be significantly higher in the presence of a palpable mass.[3] Treatment of paget's disease includes mastectomy with axillary staging or wide local excision of nipple and areola to achieve clear margins, axillary staging and radiation therapy.[4] Our patient had lesion over the whole left breast and was planned for modified radical mastectomy. The histopathologic picture was of invasive ductal carcinoma with metastatic lymph nodes. Radiotherapy department was consulted for further management of patient.

Without treatment, the skin lesions invariably spread progressively before the development of invasive breast cancer, manifesting with a palpable mass followed by lymph node and visceral metastasis. Factors of unfavorable prognosis include the presence of a palpable breast tumor, lymph node enlargement, histological type of breast cancer, and age younger than 60 years.[5,6]

CONCLUSION:

Paget's disease is a rare condition with distinct clinical entity and should always be kept in differential diagnosis while dealing with breast lesions. The surgical treatment plan must be based on clinical and radiological assessment of the patient. Postoperative adjuvant therapies should be based on the final tumor node metastasis stage.

REFERENCE 1. Cansu Karakas. Paget's disease of the breast. J Carcinog. 2011; 10: 31. | 2.Sakorafas GH, Blanchard K, Sarr MG, Farley DR. Paget's disease of the breast. | Cancer Treat Rev. 2001 Feb;27(1):9-18. | 3. Ikeda DM, Helvie MA, Frank TS, Chapel KL, Andersson IT. Paget disease of the nipple:radiologic-pathologic correlation. Radiology. 1993;189:89–94. | 4. Townsend CM., BeauchampRD., Evers BM, Mattox KL. Sabiston Textbook of Surgery, 19th edn.Elsevier, Philadelphia. pp 830,865. | 5. Kanitakis J. Mammary and extramammary Paget's disease. J Eur Acad Dermatol Venereol.2007;21:581–90. | 6. Kawase K, Dimaio DJ, Tucker SL, Buchholz TA, Ross MI, Feig BW, et al. Paget's disease of the breast: there is a role for breast-conserving therapy. Ann Surg Oncol. 2005;12:391–7. |