

Case Report

Thirty Year Old Female Presented with Nipple Destruction: A Case Report

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Abstract:

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Paget's disease of the breast is a rare type of cancer of the nipple-areola complex and that is often associated with an underlying in situ or invasive carcinoma. It is often misdiagnosed as eczema of breast and treatment is delayed. Here we present a case where a 30 year old female presented with itching ulceration and destruction of her left nipple. She was treated initially by local physicians by applying local ointments but as her condition did not improve she was admitted to department of surgery Shaheed Suhrawardy Medical College hospital where she was diagnosed as Paget's disease with infiltrating ductal cell carcinoma. She underwent modified radical mastectomy with axillary clearance and referred to oncology department for further management.

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Introduction:

Paget's disease of the breast is a malignant disease that presents itself as eroding and bleeding ulcer of the nipple. It represents an extension of a ductal breast adenocarcinoma.¹ Microscopically, typical large clear cells (Paget's cells) with pale and abundant cytoplasm and hyperchromatic nuclei with prominent nucleoli are found in the epidermal layer.² Paget's disease is more often associated with primary invasive or in situ carcinoma of the breast.³ It was first described by Sir Paget in 1874 as an eczematous lesion of the nipple associated with an underlying cancer.⁴

Paget's disease of the breast is a rare histological breast cancer, representing 1–3% of female breast cancers.⁵ It appears as an isolated affection on 11–13% of cases and is associated with an in situ or invasive glandular carcinoma on 90–100% of cases.⁵

Paget's disease of the nipple develops insidiously. Most often unilateral, it initially touches the nipple and then shows a centrifugal growth to reach the areola and then the adjacent skin. It takes the aspect of an eczema which sometimes associated with an erythema oozing. The color of the skin changes from pink to red. Retraction, ulceration, or bleeding of the nipple is possible in advanced Paget's disease. The symptoms usually reported are pruritus, burning, tingling, and pain. In 33% of cases, a palpable mass is present at the time of diagnosis.⁵ In 54% of cases, there are enlarged axillary lymph nodes.⁵

There are two main theories regarding the pathogenesis of Paget's disease: The first epidermotropic theory claims that Paget cells originating from ductal cancer cells involve chemotaxis and invasion of the epidermis of the nipple. According to the second theory, which is more accepted, Paget cells are formed from in-situ malignant keratinocytes as a result of in-situ malignant transformation or degeneration of the substantial cells.⁶

Mastectomy with or without axillary lymph node dissection has long been regarded as the standard therapy.⁷ Recent reviews have shown that conservative breast surgery combined with radiation therapy is a feasible alternative

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for patients with limited disease: long-term breast-conserving surgery would be equivalent to mastectomy in terms of overall disease-free survival.⁷ Here we present a case of Paget's disease with underlying ductal cell carcinoma admitted in the department of surgery of Shaheed Suhrawardy Medical College hospital.

Case Summary:

A 30 years old married female was admitted to department of surgery of Shaheed Suhrawardy Medical College Hospital with the complaints of Itching of her left nipple for 1 year. She also mentioned about destruction and sloughing out of her nipple 3 months back. She also gave history of anorexia and weight loss for same duration. With these complaints she visited a local physician and treated with topical ointments. But her condition did not improve. On examination her vital signs were within normal limits. On local examination the right breast appeared normal. On the left breast, nipple was distorted and there was an ulcer over the areola. (Fig-1) On palpation there was a hard lump just behind the areola about 4cm×4 cm which was mobile, hard in consistency having irregular surface and well defined margin. The central group of right sided axillary lymph nodes were also enlarged. The largest one was measuring about 3cm×2 cm, firm, and mobile. Other systemic examination revealed normal findings. Her



Fig1: showing destruction of nipple and ulceration.

Complete blood count showed normal haemoglobin level with raised ESR (55 in 1st hr.). Ultrasonogram of both breast revealed left sided suspected malignant lesion with axillary lymphadenopathy. Mammogram of both breasts showed similar findings. (Fig-2 and FIG-3) FNAC result showed suggestive of inflammatory lesion. So trucut biopsy was done from the lump and biopsy was also taken from the ulcerated lesion over areola. Histopathology report showed Paget's disease (Fig-4) with infiltrating ductal cell

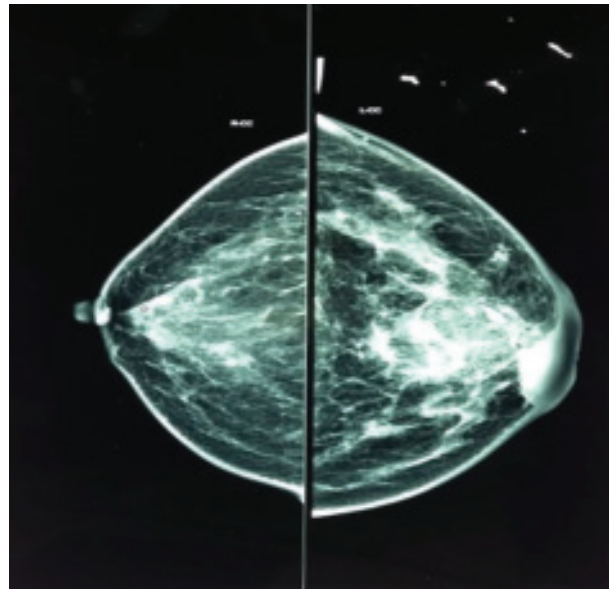


Fig-2: mammogram of both breasts

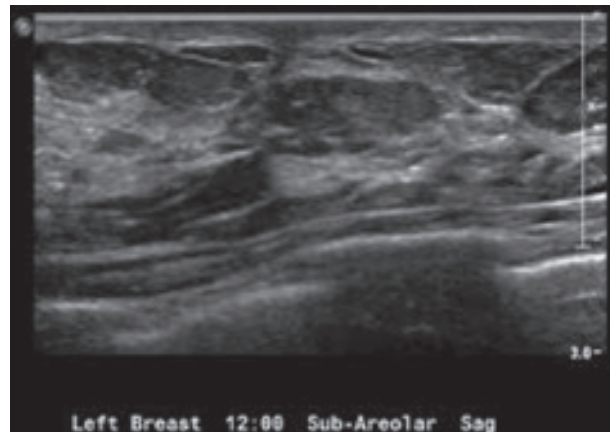


Fig-3: Ultrasonogram of left breast

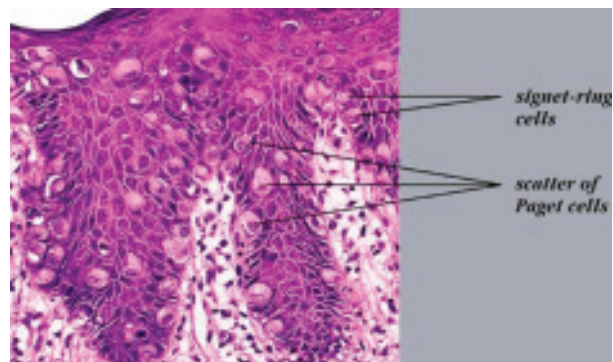


Fig-4: Histopathological view of Paget's disease

carcinoma. Her other investigations including renal function, liver function, blood sugar, Chest X-ray and ECG demonstrated normal findings. After consultation from oncology department she underwent modified radical mastectomy with axillary clearance. Her post-operative period was uneventful. She was referred to oncology

department on 10th post-operative day for further management.

Discussion:

Paget's disease of the breast is a rare disease that usually occurs in the form of an eczema-like lesion on the nipple. Most patients initially present with pain, redness, burning or pruritus of the nipple, which is likely indicating the presence of eczema and is often treated as dermatitis or eczema, leading to the possibility of the diagnosis of being Paget's disease missed. Topical steroids given for eczema diagnosis can cause some improvement in the inflammatory component of the disease and mask the underlying pathology for a while. Other diseases that have differential diagnosis are benign intraductal papilloma, basal cell carcinoma, squamous cell carcinoma, superficial spreading malignant melanoma, psoriasis, contact dermatitis, syphilis and Bowen's disease.⁸ In more than 90% of cases, it is associated with underlying in situ or invasive breast carcinoma.⁹ Due to the high incidence of multifocal involvement, conventional treatment has been mastectomy with or without axillary lymph node surgery.

Histopathological examination of the lesion reveals round, oval atypical Paget cells with a large, pleomorphic, hyperchromatic nucleus and a clear cytoplasm. These cells can be observed intraepidermally.¹⁰ In our case, histopathological report showed similar results.

When a female exhibits an eczematous change, ulceration on her nipple, palpably enlarged axillary lymph nodes, a palpable breast lump and nipple discharge, bleeding, pruritus and induration, the first steps for evaluating a suspicious breast lesion are ultrasonography and mammography. Suspicious micro-calcifications can be found by breast mammography; on ultrasonography, invasive cancers are typically solid, and all solid lesions require biopsy.¹¹ When there are no palpable masses or mammography findings, MRI may play an important role in selecting suspicious candidates.¹² Any suspicious lesion requires biopsy or exfoliative cytology to confirm the diagnosis before operation. In our case as FNAC was non-conclusive we performed biopsy which confirmed the diagnosis.

Because of the high incidence of multifocal and multicentric cancer, the treatment of Paget's disease has traditionally been mastectomy.¹³ Breast-conserving treatments could be used in selected female patients. In our case, as there was underlying ductal cell carcinoma with Paget's disease, the patient underwent modified radical mastectomy with axillary clearance.

Conclusion:

Paget's disease of the breast is a rare cancer. It must be distinguished from an eczematous involvement of the nipple, not responding to a local treatment. The recommendations of the treatment are limited by the absence of randomized prospective trials comparing mastectomy to conservative surgery or by comparing various options for conservative surgery in patients with Paget's disease of the breast. The prognosis depends on the presence of a palpable mass and the invasiveness of the underlying cancer. So we recommend early diagnosis and prompt management for the better prognosis of patient.

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