

# Paget's Disease, Breast

Paget's disease of the breast, also known as Paget's disease of the nipple, is a rare form of breast cancer that primarily affects the nipple and areola. It was first described by Sir James Paget in 1874. This condition is often marked by a persistent, eczematous lesion on the nipple and surrounding areola, which may resemble inflammatory skin conditions such as eczema or dermatitis. However, it is important to differentiate Paget's disease from eczema, as Paget's disease typically affects only one breast, whereas eczema usually impacts both breasts. Recognizing these differences is crucial for appropriate diagnosis and treatment.

## Pathophysiology and Clinical Features

Paget's disease of the breast is often associated with underlying breast cancer, especially invasive ductal carcinoma (IDC) or ductal carcinoma in situ (DCIS). The disease originates in the milk ducts, where cancer cells spread to the nipple, resulting in characteristic skin changes. The malignant cells disrupt the normal epidermis of the nipple and areola, leading to inflammation and the formation of visible lesions. The condition usually presents unilaterally and includes the following features:

- ***Nipple and Areolar Changes:*** The skin on the nipple and areola becomes dry, scaly, red, and may resemble eczema or dermatitis. It can also become crusty, fissured, or show a dimpled appearance (peau d'orange). In some cases, the nipple may become inverted.
- ***Itchiness and Discomfort:*** Patients often report itching and tenderness in the affected area, with some experiencing nipple discharge that can be clear, bloody, or yellowish.
- ***Fluctuating Symptoms:*** The lesions can fluctuate in severity, improving and worsening over time. This variability may lead to a delayed diagnosis, as symptoms might be mistaken for benign conditions like eczema or dermatitis.
- ***Progression:*** Without timely treatment, Paget's disease can progress to involve the underlying breast tissue, leading to ulceration, widespread skin changes, and potentially affecting deeper breast structures.

## Diagnosis

The diagnosis of Paget's disease of the breast is primarily based on clinical examination, but confirmation requires histopathological analysis. A biopsy of the affected nipple or areolar skin is essential to identify the presence of malignant cells. Since Paget's disease is almost always associated with underlying breast cancer, particularly invasive ductal carcinoma (IDC) or ductal carcinoma in situ (DCIS), mammography is commonly used to screen for these conditions.

Additionally, fluid obtained from nipple discharge (if present) can be analyzed cytologically to help confirm the diagnosis and assess the presence of cancer cells. Early diagnosis through these methods is crucial for effective treatment and management.

## Treatment Options

The treatment of Paget's disease of the breast generally follows the principles used in the management of breast cancer. The primary goal is to remove cancerous tissue and manage any underlying malignancy. Treatment options include:

- **Surgical Intervention:** Surgery is the cornerstone of treatment. For localized Paget's disease, a wide excision of the affected tissue, including the nipple and areola, may be performed. In more extensive cases, a total mastectomy may be necessary, particularly if there is significant underlying invasive carcinoma.
- **Radiation Therapy:** Radiation therapy is often used after surgery, especially when breast-conserving surgery is performed. It may target any residual cancer cells or treat positive surgical margins to reduce the risk of recurrence.
- **Chemotherapy and Hormonal Therapy:** Chemotherapy is indicated for more advanced cases or if metastasis is present, particularly in patients with invasive ductal carcinoma. Hormonal therapy, such as tamoxifen, may be recommended for patients with hormone receptor-positive tumors.
- **Targeted Therapies:** In some cases, especially when the tumor is HER2-positive, targeted therapies like trastuzumab may be considered. These therapies work to inhibit cancer growth and can improve survival outcomes.

## Prognosis and Outcome

The prognosis of Paget's disease of the breast largely depends on the stage at diagnosis and the presence of underlying breast cancer. If detected early and treated effectively, particularly before significant spread to other tissues, the prognosis can be favorable. In cases where Paget's disease is associated with early-stage breast cancer or ductal carcinoma in situ (DCIS), patients typically have a better outcome, with high survival rates.

However, if Paget's disease is associated with more advanced invasive carcinoma or metastasis to lymph nodes, the prognosis becomes less favorable. The spread of cancer beyond the breast tissue or to distant organs is associated with a poorer survival rate. The identification and treatment of underlying invasive breast cancer are crucial, as they directly impact treatment decisions and overall survival outcomes. Early detection, effective treatment, and close monitoring are key to improving the prognosis for individuals with Paget's disease of the breast.

## Conclusion

Paget's disease of the breast is a rare but significant condition that primarily affects the nipple and areola, often linked to underlying breast cancer. Early diagnosis through biopsy and imaging is crucial for effective treatment. Management typically involves surgical excision, radiation therapy, chemotherapy, and targeted therapies, with treatment tailored to the extent and stage of the underlying malignancy. The prognosis largely depends on how early the disease is detected and the presence of invasive cancer. Given the fluctuating nature of the skin lesions, healthcare providers must maintain a high index of suspicion for Paget's disease in patients with nipple and areolar changes, ensuring timely diagnosis and intervention.

## References

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