

Paget Disease of the Breast

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The Operative Review of Surgery. 2023; 1:17-19.

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General Information

Definition

- A Rare Type of Breast Cancer with Paget Cells Involving the Nipple-Areola Complex
- Also Known as Paget's Disease of the Nipple or Mammary Paget Disease



Pathophysiology

- *Epidermotropic Theory*: Paget's Disease Derives from Underlying Breast Cancer with Malignant Ductal Cells Migrating Through the Ductal System to the Nipple ²
 - *By Far the More Commonly Accepted Theory for the Majority of Cases
- *Transformation Theory*: Paget's Disease Derives from Malignant Transformation of Nipple-Areolar Keratinocytes Independent of Underlying Disease ²
- *Dual Origin Theory*: Depending on Circumstances, Paget's Disease May Originate from Either Ductal Cancer Cells or Malignant Transformation of Keratinocytes ²

Underlying Malignancy

- Paget's Indicates an Underlying **Ductal Carcinoma In Situ (DCIS)** or **Invasive Ductal Carcinoma (IDC)** in 80-90% of Cases ³
- 1-4% of All Breast Cancer Patients Also Have Paget's Disease of the Breast ⁴
- About 50% of Cases Have are Associated with a Palpable Mass ⁵
 - Mass is Most Common Centrally Located but Can Be Seen in Periphery with Multifocal or Multicentric Disease ⁵
- May Be Seen as a Form of Local Recurrence After Nipple-Sparing Mastectomy ⁶

Epidemiology

- Most Common in Postmenopausal Women Aged 50-60 ⁵
- Reported Age Range: 20-90 Years ⁷
- Majority in Women (97.9%) But Can Affect Men (2.1%) ⁷

Presentation ^{3,8}

- **Eczematoid Scaling of the Nipple-Areola Complex**
- Pain, Itching, & Burning
- May Have Vesicles or Ulcerations
- Copious Clear-Yellow Exudate – Later Feature ⁹
- Flattened or Inverted Nipple – Later Feature ⁹
- *Often Misdiagnosed as Eczema, Psoriasis, or Dermatitis ^{3,10}

Diagnosis & Treatment

Diagnosis

- Should Undergo a Bilateral Diagnostic Mammogram to **Evaluate Underlying Lesion** ^{8,9}
 - May Also Consider an MRI to Evaluate for More Occult Lesions that May Be Missed on Mammogram ¹¹
- Definitive Diagnosis: **Full Thickness (Punch) Skin Biopsy of the Nipple** ⁹
 - May Also require Core Needle Biopsy (CNB) to Diagnose the Primary Lesion
- Pathology: Paget Cells are Malignant Adenocarcinoma Cells within the Epidermis of the Nipple
 - Large, Pale-Staining Cells with Round or Oval Nuclei & Large Nucleoli Located Between the Normal Keratinocytes ¹²
 - Does Not Invade Through the Dermal Basement Membrane ¹²

Staging

- Classified as Carcinoma In Situ (Tis) ¹³
- Paget's Disease of the Breast Does Not Change the Stage of an Underlying Malignancy ¹³

Treatment

- General Treatment: **Simple Mastectomy & Sentinel Lymph Node Biopsy (SLNB)**
 - All Patients Require Resection of the Nipple-Areola Complex ⁹
- May Consider Breast-Conserving Treatment (BCT) in Select Cases ¹⁴
 - Includes: Wide Local Excision (WLE) of the Nipple, Lumpectomy, SNLB, and Whole Breast Radiation
 - Consider Preoperative Evaluation with MRI to Rule Out Occult Multicentric Disease ¹⁵
 - Exact Indications for BCT are Not Yet Well Defined
- 5-Year Survival Rate After Surgery: 82.5% ⁷

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