Paget Disease of Breast

**PRESENTATION**

- Clinical breast exam
- Medical photography
- Diagnostic bilateral mammography, ipsilateral ultrasound of nodal basins and ultrasound evaluation with focus of nipple/retro-areolar breast
- Full thickness skin biopsy of involved nipple areola complex (NAC)
- Lifestyle risk assessment

**INITIAL EVALUATION**

- NAC biopsy negative and imaging negative
  - Clinical follow-up
  - Re-biopsy if not healing
- NAC biopsy positive and imaging negative
  - MRI breast with and without contrast and biopsy of MRI-detected lesion
- NAC biopsy positive and imaging positive
  - Core biopsy of breast lesion
  - Consider MRI breast with and without contrast if patient desires breast conserving surgery

**FURTHER EVALUATION AND TREATMENT**

- Individualized care based on clinical indications and risk assessment
- Consider Dermatology consult to evaluate for the causes of skin symptoms and findings

**Note:** Consider clinical trials as treatment options for eligible patients.

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1 Nipple or areola changes such as eczema, ulceration, bleeding, itching
2 For medical photography, refer to Photographs – HIPAA Authorizations General Reference Tool (ATT1597)
3 See Physical Activity, Nutrition, and Tobacco Cessation algorithms; ongoing reassessment of lifestyle risks should be a part of routine clinical practice
4 For surveillance guidelines, see Breast Cancer Screening algorithm
5 To assess the extent of disease or confirm additional disease, consider MRI breast (see Breast Cancer – Invasive algorithm)
**Paget Disease of Breast**

**TREATMENT**

- No breast lesion and NAC Paget without invasion
  - Mastectomy to include NAC with axillary staging or Complete excision of NAC without axillary staging
  - Appropriate systemic adjuvant and/or radiation therapy
  - Surveillance

- No breast lesion and NAC Paget with invasion
  - Mastectomy to include NAC with axillary staging or Complete excision of NAC with axillary staging
  - Appropriate systemic adjuvant and/or radiation therapy
  - Surveillance

- Breast Ductal Carcinoma In Situ (DCIS) and NAC Paget
  - Mastectomy¹ to include NAC with axillary staging or Breast conserving surgery: complete excision of the nipple areolar complex and excision of the breast tumor⁴ followed by whole breast radiation⁵
  - Appropriate systemic adjuvant and/or radiation therapy
  - Surveillance

- Breast invasive¹ and NAC Paget
  - Mastectomy³ to include NAC with axillary staging or Overall management is based on the extent and stage of the underlying breast cancer. Breast conserving surgery: complete excision of the NAC and excision of the breast tumor⁴ followed by whole breast radiation⁵
  - Appropriate systemic adjuvant and/or radiation therapy
  - Surveillance

**SURVEILLANCE**

- Surveillance²

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¹ Depending on the stage and pathology, see either Breast Cancer – Ductal Carcinoma in Situ (DCIS) algorithm or Breast Cancer – Invasive algorithm as appropriate

² Paget Disease with invasion but without underlying carcinoma, see Breast Cancer – Invasive algorithm

³ Paget disease with invasion and with concurrent DCIS, see Breast Cancer – Invasive algorithm

⁴ Breast Cancer – Invasive algorithm as appropriate

⁵ Mastectomy is always an option with any manifestation of Paget disease

⁶ The NAC and the peripheral cancer can be excised as separate surgical specimens and do not need to be removed as one contiguous mass

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Disclaimer: This algorithm has been developed for MD Anderson using a multidisciplinary approach considering circumstances particular to MD Anderson’s specific patient population, services and structure, and clinical information. This is not intended to replace the independent medical or professional judgment of physicians or other health care providers in the context of individual clinical circumstances to determine a patient’s care. This algorithm should not be used to treat pregnant women.

Note: Consider clinical trials as treatment options for eligible patients.

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SUGGESTED READINGS


This practice consensus algorithm is based on majority expert opinion of the Breast Faculty at the University of Texas MD Anderson Cancer Center. It was developed using a multidisciplinary approach that included input from the following:

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