

Patient Diagnosis Resource for PAGET'S DISEASE OF THE NIPPLE

Your Diagnosis

Your doctor has determined that you have Paget's disease of the nipple, a rare form of breast cancer that accounts for only about 1 to 4% of all breast cancer cases.

About the Condition

The breast is comprised of 2 main types of tissue — glandular and supportive. The glandular portion includes the lobules, which produce milk in women who are breastfeeding, and the ducts, which carry milk from the lobules to the nipple. The supportive portion includes the fibrous connective tissue and fatty tissue that determine the size and shape of the breast. Any of the tissues of the breast can experience symptom-causing changes, which may be either benign or cancerous.

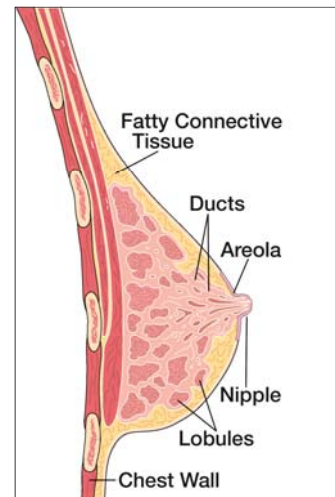
Generally Paget's disease of the nipple starts in the ducts and extends to the skin of the nipple and then to the areola and adjacent skin. Usually one breast is affected, but it can affect both breasts.

About 88% of patients with Paget's disease of the nipple have underlying invasive ductal carcinoma (IDC), which is the most common type of invasive breast cancer. IDC occurs when cancerous (malignant) cells form in a milk duct and spread outside the duct into nearby breast tissue forming a lump, or tumor.

Some 9% of those with Paget's disease of the nipple have underlying ductal carcinoma in situ (DCIS), an early stage of breast cancer that forms inside a milk duct. DCIS is the most common breast cancer that is non-invasive, or contained within the area where it began.

In 3% of cases the cancer is confined to the nipple area, with no underlying invasive ductal carcinoma or ductal carcinoma in situ.

The outlook for patients with Paget's disease of the nipple varies depending on factors such as the presence or absence of a breast lump and IDC or DCIS. Your doctor may want to perform one or more tests to help determine if the cancer has spread beyond the breast, which could include a mammogram (breast X-ray), MRI scan, CT scan, lymph node biopsy, bone scan or PET scan. Cancer that is confined within the breast is the most manageable and curable. If malignant cells extend into the lymph nodes and beyond to other areas of the body, the treatment plan will be more complex and the cancer will be more difficult to manage. Cases of advanced breast cancer are referred to as metastatic breast cancer.



Treatment Options

Deciding on a treatment plan for your breast cancer can be complex and depend upon a variety of factors such as your age, general health condition, stage of cancer and personal preferences. Sometimes more than one type of therapy may be used.

The following treatment possibilities are available:

Lumpectomy – A lumpectomy is breast-conserving surgery that removes only the nipple and areola and a surrounding area of normal tissue. This can be an option in the rare cases where there is no evidence of underlying cancer, when only the nipple and adjacent area are found to be cancerous.

Mastectomy – Mastectomy, or surgery to remove the breast, may be performed with or without reconstructive surgery. A total mastectomy removes the entire breast but none of the axillary (underarm) lymph nodes and is an option in cases where the underlying cancer is not invasive. In cases of invasive breast cancer or very extensive ductal carcinoma in situ, a modified radical mastectomy may be performed, which removes the entire breast and some of the axillary lymph nodes. Patients considering breast reconstruction should consult with a plastic surgeon who is an expert in the procedure before having a mastectomy.

Radiation Therapy – Lumpectomy is usually followed by radiation therapy, which can be delivered externally or internally. In external beam radiation, a high energy X-ray machine is used to direct radiation at the tumor. Internal radiation therapy uses small radioactive implants placed directly into the breast.

Hormone Therapy – Hormone therapy is only recommended for women whose cancer cells have certain hormone receptors as determined by laboratory testing. It helps cancer shrink and grow more slowly by keeping the malignant cells from exposure to certain hormones such as estrogen and progesterone. Methods used include drugs such as tamoxifen to block the production and effect of the hormones and the removal of the ovaries, the main site of estrogen production.

Chemotherapy – The use of anti-cancer drugs, or chemotherapy, provides a way to slow tumor growth and reduce pain for patients whose cancer has spread. Chemotherapy can work in one of two ways: by keeping the cancer cells from multiplying or by causing the cancer cells to die.

You may also want to talk with your doctor about participating in a clinical trial, which helps physicians learn about new treatments and better ways to use established treatments.

What You Can Do

You can choose to take an active role in your health and well-being. Learn as much as you can about your condition and have a list of questions ready each time you meet with your doctor. Join a cancer support group, and talk with your family, friends, clergy person or counselor as you feel comfortable. Also, be sure to get enough sleep every night.

Other steps you can take to maximize your health include eating a low-fat diet high in fruits and vegetables, avoiding the use of tobacco, limiting consumption of alcohol and red meat, taking part in some form of exercise and maintaining a healthy body weight.

Additional Resources

American Cancer Society, 800.227.2345, www.cancer.org

National Cancer Institute, 800.422.6237, www.cancer.gov

Susan G. Komen Breast Cancer Foundation, 800.462.9273, www.komen.org

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