A 44-year-old female has a 3-month history of a pruritic and erythematous plaque on the left nipple and areola. The patient has been using mometasone furoate cream which provided inadequate relief.

What is your diagnosis?

Mammary Paget disease (PD) is a less common form of breast cancer which involves the nipple-areola complex and occurs almost exclusively in females. Male breast involvement is rarely reported. Erythema, skin thickening, pruritus, burning sensation, inversion of the nipple, ulceration, serosanguineous nipple discharge are common clinical symptoms. Lesion size ranges from 3 mm to 15 cm in diameter; the mean size is 2.8 cm in diameter. A similar disease involving the skin of external genitalia is known as extramammary Paget disease.

Nipple changes are associated with an underlying carcinoma of the breast in over 98% of patients with mammary PD. Up to 2/3 of patients have a palpable breast...
tumor. Nearly all cases have unilateral involvement, but bilateral mammary PD has occasionally been reported. Rare cases of female patients with PD of supernumerary nipples have been reported.

Paget cells may derive from luminal lactiferous ductal epithelium of the breast tissue. Malignant Paget cells infiltrate and proliferate in the epidermis, causing thickening of the nipple and the areolar skin.

The exact frequency of mammary PD is unclear. Approximately 1-4% of female breast carcinoma are associated with PD of the nipple-areola complex. Almost 100% of mammary PD are associated with either in situ (10%) or infiltrating cancer (90%). The average age of diagnosis is 53–59 years old.

It is not uncommon that symptoms are present for 6 months or more before the underlying breast cancer is diagnosed. A biopsy including the dermal and subcutaneous tissue should be performed on all suspicious lesions of the nipple-areola complex for accurate diagnosis and treatment of mammary PD. Even though one half of breast carcinomas are positive for these hormone receptors, tumorous Paget cells are negative for estrogen and progesterone receptor sites.

Mammary PD has been classified into 4 clinical stages:

Stage 0—Lesion confined to the epidermis, without underlying in situ ductal carcinoma of the breast

Stage 1—Associated with in situ ductal carcinoma just beneath the nipple

Stage 2—Associated with extensive in situ ductal carcinoma

Stage 3—Associated with invasive ductal carcinoma

Of all patients with mammary PD, 40-50% have either stage 1 or 2 disease and they have no palpable breast tumor. A palpable breast tumor must be present in stage 3; more than half of patients have coexisting axillary lymph node involvement.

The first line treatment of mammary PD is mastectomy (radical or modified) and lymph node clearance for patients with a palpable mass and underlying invasive breast carcinoma. Up to 2/3 of patients are reported to have metastasis to the axillary lymph nodes. Conservative treatment with preservation of the nipple-areola complex results in a higher rate of recurrence. Radiation therapy

Figure 1: Mammary Paget disease
Mammary Paget disease (PD)

Gross Appearance

- Ulceration
- Inverted nipple
- Erythema

Mean lesion size = 2.8 cm diameter

Clinical Staging of Mammary PD

**Stage 0**
- Lesion confined to epidermis of nipple and areola

**Stage 1**
- In situ ductal carcinoma just beneath the nipple

**Stage 2**
- Extensive in situ ductal carcinoma

**Stage 3**
- Invasive ductal carcinoma

Lactiferous duct
A Pruritic Rash

**SUMMARY OF KEY POINTS**

Erythema, skin thickening, pruritus, burning sensation, inversion of the nipple, ulceration, serosanguineous nipple discharge are common clinical symptoms of mammary Paget disease (PD).

Paget cells may derive from luminal lactiferous ductal epithelium of the breast tissue. Malignant Paget cells infiltrate and proliferate in the epidermis, causing thickening of the nipple and the areolar skin.

alone does not always adequately control occult breast cancer, but it may be used for patients who refuse mastectomy or those who are not suitable for surgery. Ami-nolevulinic acid photodynamic therapy has been suggested as adjunctive therapy for selected patients by inducing local cytotoxicity through a photodynamic reaction. This treatment may be appropriate for low-risk malignant cells and improve morbidity.6

The prognosis of mammary PD is determined by the disease stage and is similar to that of other types of breast cancer. The prognosis is worse with the presence of a palpable breast mass. For patients with PD without a palpable breast tumor (prior to surgery), the reported survival rate is 92–94% at 5 years and 82–91% at 10 years. For patients with an identifiable breast tumor, the survival rate is reduced to 38–40% at 5 years and 22–33% at 10 years. The death rate for metastatic breast cancer in patients with mammary PD and underlying cancer is 61.3%.1

**References**


All of the tables and photos are original.
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