Mammary Paget Disease

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A 45-year-old woman presented with a 4-month history of a 3.5-cm pruritic scaly plaque with an irregular border and central ulceration (image A, arrow) of her left nipple-areola complex. Although the patient had no other relevant medical history and no palpable breast masses or nodules, mammary Paget disease was suspected. A wedge biopsy and histopathologic examination (image B; hematoxylin-eosin stain; original magnification, ×40) revealed intraepidermal Paget cells (arrows) with moderate amounts of pale cytoplasm and enlarged pleomorphic nuclei. Mammographic microcalcification patterns (not pictured) suggested subareolar ductal carcinoma in situ. The patient underwent a partial mastectomy with removal of the nipple-areola complex and underlying microcalcifications followed by sentinel node biopsy, which was negative for tumor cells. The patient tolerated both the partial mastectomy and sentinel node biopsy well and is currently undergoing radiation therapy.

Mammary Paget disease is characterized by intraepidermal infiltration by malignant glandular epithelial cells. The age-adjusted incidence decreased from 1.31/100,000 woman-years in 1988 to 0.64/100,000 woman-years in 2002. In the United States, 82% to 87% of Paget disease cases include underlying ductal carcinoma in situ or invasive ductal carcinoma, but breast masses are palpable in only 14% to 44% of cases. Physicians should maintain a high level of suspicion for nipple-areolar abnormalities, as the superficial lesion of mammary Paget disease may be the only sign of an underlying neoplasm.

References