

## Answer to Dermacase continued from page 33

### 2. Paget disease

Paget disease (PD) is rare and accounts for approximately 1% to 3% of all patients with breast cancer.<sup>1,2</sup> It typically occurs in postmenopausal women, with a peak incidence among those who are 50 to 60 years of age.<sup>3</sup> Men can also develop PD of the breast, but it is extremely rare.

Progression of PD is slow. It typically starts as a scaly erythematous patch on the nipple, then spreads to the areola.<sup>3</sup> It is well demarcated from the surrounding normal skin. Early symptoms can include itchiness and burning sensations. The eczematous change can temporarily resolve with corticosteroid treatment. In addition, tiny vesicles can also appear and disappear on the nipple area. This adds to a delay in diagnosis. Typically, 10 to 12 months can pass before PD is correctly diagnosed.<sup>3</sup> In the latter stages, PD can ulcerate and destroy the nipple or areolar complex. Bloody discharge is also not uncommon at this stage.

There are typically 3 possible clinical patterns of the disease: changes in the nipple or areolar complex only, changes in the nipple or areolar complex with an underlying breast mass, or clinical presentation of a breast mass with histologic confirmation of PD.<sup>3</sup> Prognosis and treatment are dependent on the presence or absence of palpable masses or axillary nodes and underlying invasive carcinoma.

### Treatment

Primary treatment of PD is still surgical.<sup>4</sup> However, due to conflicting theories of how PD develops, there is much controversy with regard to radical versus conservative surgical treatment. In the epidermotropic ductal theory, it is believed that the cancer cells associated with PD originate from the subareolar ducts, migrating from its basal membrane to the nipple epidermis.<sup>1,3</sup> The in situ malignant transformation theory suggests that cancer cells originate from the epidermis itself.<sup>1,3</sup> If the epidermotropic ductal theory is correct, lumpectomy or mastectomy are the treatments of choice. If the in situ theory is correct, skin excision alone would be the most appropriate therapy for limited disease.


In patients with only nipple or areolar complex involvement, some experts recommend skin excision with consideration of adjunct radiation therapy.<sup>5</sup> There have been some reports of using radiation therapy alone, with varying results. At present, radiation therapy alone is not considered a standard method of treatment.<sup>4</sup>

In patients with a palpable mass and negative skin margins, lumpectomy with adjunctive radiation is recommended. Both nipple or areolar complex and



unifocal disease should be followed by regular mammography. Among patients choosing not to have post-operative radiation therapy, the 5-year recurrence rate is 28% versus 5% among those who do have radiation therapy. Mastectomy should be reserved in those with relapses.<sup>4</sup> In patients with a palpable mass and involved margins, complete mastectomy is recommended.

The 5-year prognosis for patients without breast masses is 85% versus 32% with breast masses.<sup>3</sup> The 10-year survival rate for node-negative versus node-positive individuals is 79% versus 28%, respectively.<sup>3</sup> Men have a worse prognosis, with a reported 5-year survival rate of 20% to 30%.<sup>5</sup>

Paget disease of the breast should be strongly considered when any areolar or nipple lesion fails to heal with topical steroid therapy. Most lesions would clear within a month or so. A high index of suspicion is key to diagnosis. 

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### Competing interests

None declared

### References

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