

Mammary Paget's Disease Presenting as an Annular Plaque

Dear Editor,

Mammary Paget's disease (MPD) is an adenocarcinoma localized within the epidermis of the nipple and/or the areola of the breast, and it is as a rule associated with a carcinoma of the underlying lactiferous ducts, where it usually starts. MPD is relatively rare, observed in 0.7-4.3% of all breast cancers (1). We present a patient with MPD and atypical clinical finding as an annular plaque.

A 74-year-old Japanese woman with a past medical history of hypothyroidism presented with a 6-month history of an itching plaque on the left areola. The patient had been treated with the application of topical steroids for a duration of approximately 5 months, and showed no clinical improvement. Physical examination showed a pink plaque encircling the nipple on the left areola (Figure 1, a). The right nipple and areola appeared normal (Figure 1, b). No palpable

masses were detected within either breast. A 3.5 mm punch biopsy of the skin at the 6 o'clock position of the left areola was performed. Histological examination showed single and small aggregations of atypical cells with large hyperchromatic nuclei and pale-staining, ample cytoplasm throughout the epidermis. There was a lymphocytic infiltration in the dermis (Figure 1, c). Immunohistochemical studies were positive for CK7 and negative for S-100 and HMB45. With the diagnosis of MPD, the patient underwent a partial mastectomy of the left breast center area, consisting of surgical excision of the left nipple, the adjacent surrounding areolar skin, and subcutaneous tissues. Subsequently, radiation therapy for the residual breast was prepared.

As has been described in detail by Kanitakis, the skin lesion develops insidiously as a scaly, fissured, or

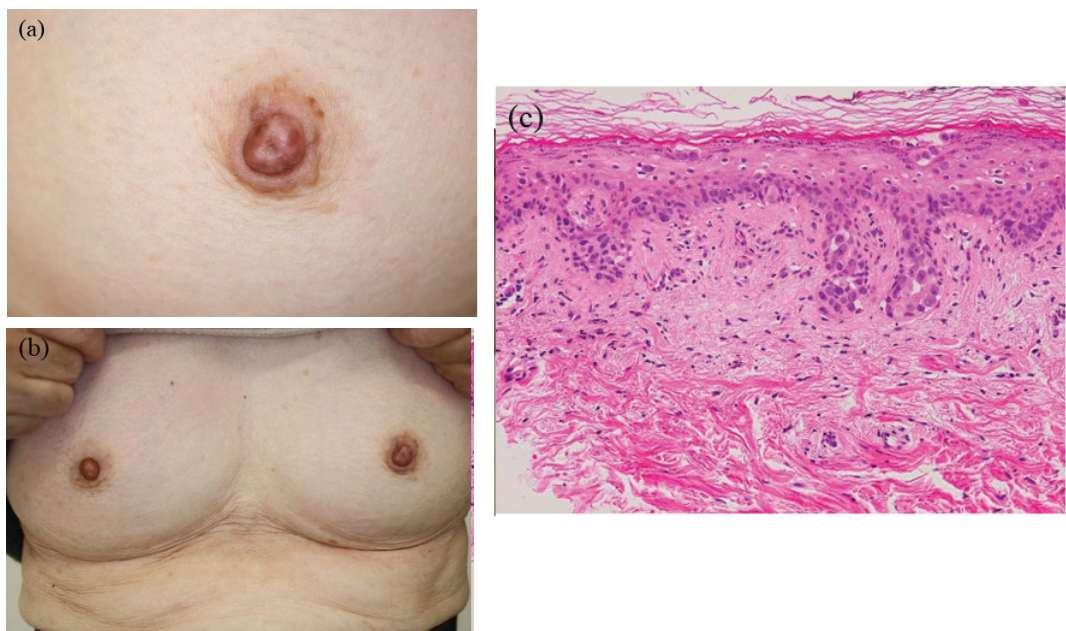


Figure 1. Pink plaque encircling the nipple on the left areola. (a) The right nipple and areola appeared normal. (b) Histopathological aspect of the lesion showing round cells in the epidermis with an ample, light-stained cytoplasm (hematoxylin and eosin stain, $\times 200$) (c).

oozing erythema of the nipple and, more rarely, the areola. Advanced lesions present as a well-demarcated, round, ovoid, or polycyclic eczema-like plaque with a pink or red hue. It is occasionally slightly infiltrated and has an erosive, oozing, scaly, or crusted surface. The lesions are almost invariably unilateral, showing centrifugal spread. Retraction or ulceration of the nipple are often noted (1). The present case exhibited a very rare clinical finding of a plaque encircling the nipple, which has not been reported previously. It was initially difficult to establish the diagnosis of MPD, and biopsy was needed to obtain a definitive diagnosis.

Differential diagnosis of MPD comprises eczema as atopic dermatitis or contact dermatitis, erosive adenomatosis, and malignant skin condition such as Bowen's disease, superficial basal cell carcinoma, or superficially spreading melanoma. As in the present case, individuals presenting with an annular plaque are often considered to have sebaceous hyperplasia. Sebaceous hyperplasia is a common, benign skin condition involving hypertrophy of the sebaceous glands, common in middle-aged or older adults (2). These lesions can be single or multiple and manifest as yellow, soft, small papules. These papules are occasionally seen around the nipple, forming an annular plaque. In general, sebaceous hyperplasia is described as yellow-colored papules among Caucasians. However, caution is needed, since it is characterized by skin-colored papules among some Asians.

In the present case, some pigmentation (2 to 3 mm in diameter) was observed on the left nipple. Pigmented MPD have been reported, and the mechanism underlying the pigmentation is not yet fully understood, but it has been proposed that Paget cells may release melanocytic chemoattractants or basic fibroblast growth factors that stimulate the proliferation of melanocytes within the tumor nests (3). The possibility of physiological pigmentation cannot be

ruled out in the present case; on the other hand, the possibility of pigmented MPD cannot be ruled out either, since no pigmentation was observed on the right nipple.

References:

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