Well-demarcated erythematous plaque on the arm

A 67-year-old Caucasian man presented with an asymptomatic, well-defined, irregular shaped, erythematous scaly plaque on the left upper arm for five years [Figure 1]. There was no cervical or axillary lymphadenopathy. A skin punch biopsy was performed from the plaque.

Question

What is your diagnosis?

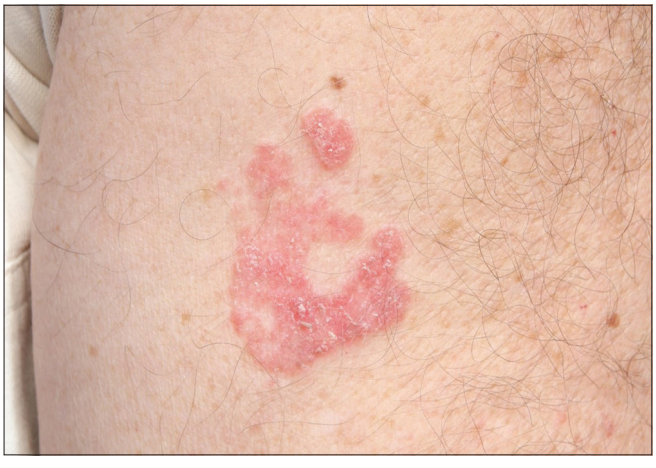

Figure 1: An irregular shaped erythematous plaque with scaling, on the left upper arm

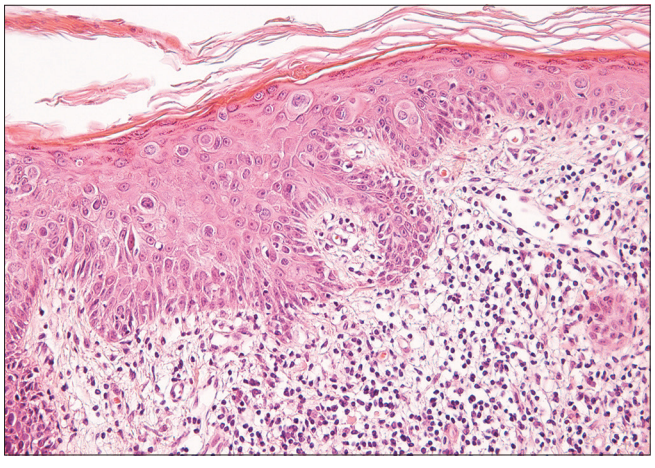

Figure 2a: Histological examination showed hyperkeratosis, acanthosis and atypical large pale cells with abundant vacuolated cytoplasm in the epidermis (H and E, ×200)

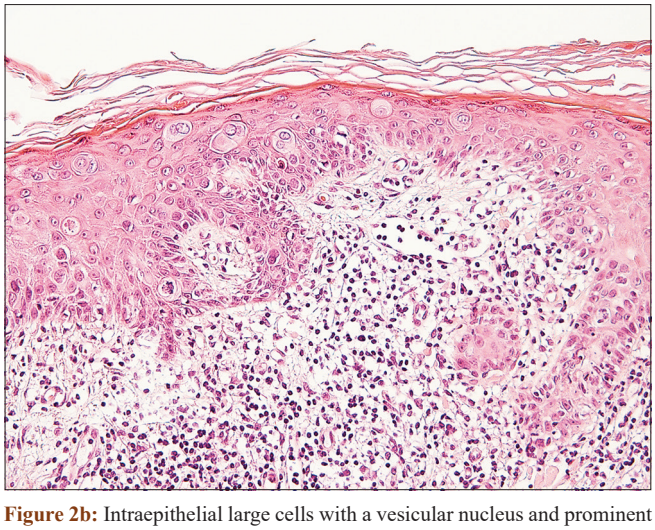

Figure 2b: Intraepithelial large cells with a vesicular nucleus and prominent nucleolus (H and E, ×400)

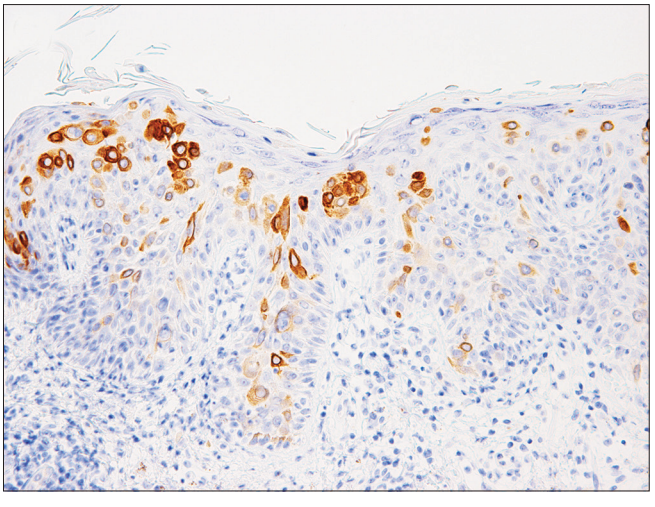

Figure 3a: Tumor cells were positive for cytokeratin 7 (immunohistochemistry, ×200)

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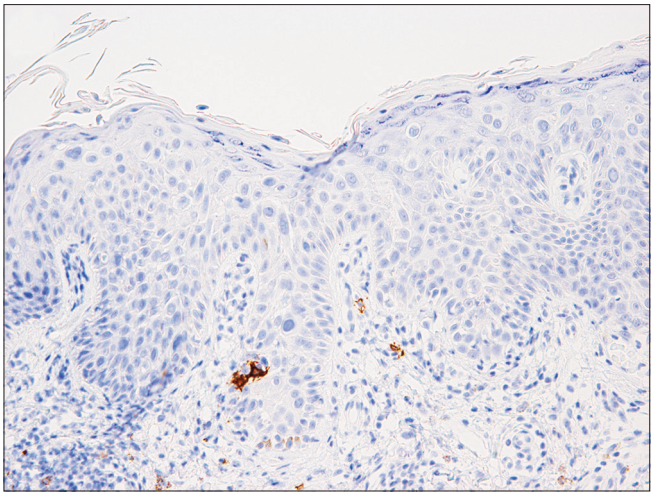

Figure 3b: Tumor cells were negative for cytokeratin 20 (immunohistochemistry, ×200)

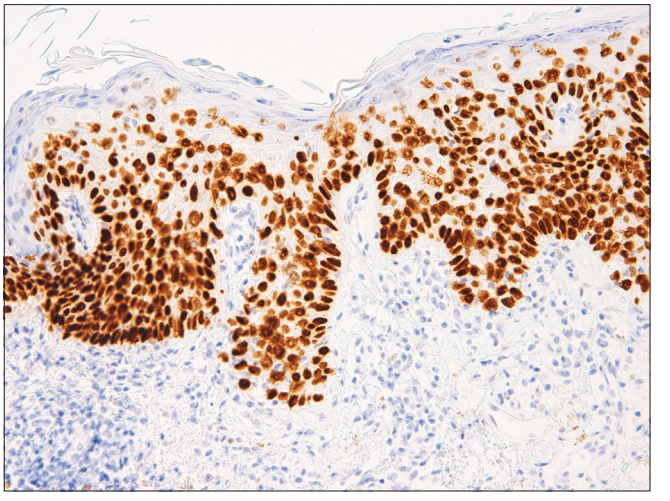

Figure 3c: Tumor cells were negative for P63 whereas normal epidermal cells were positive (immunohistochemistry, ×200)

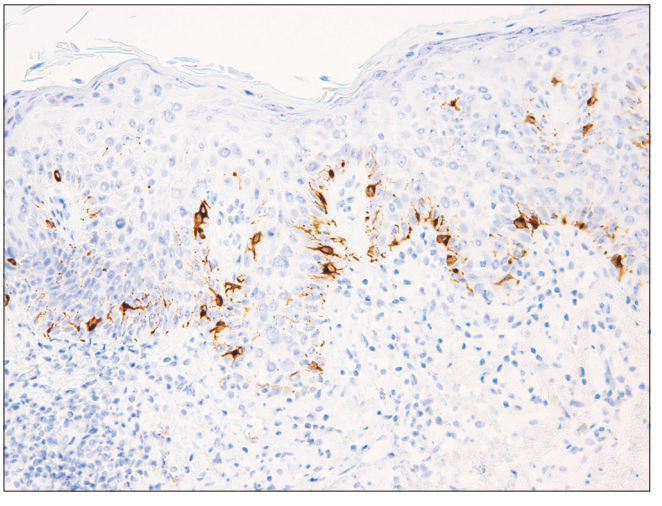

Figure 3d: Tumor cells were negative for Melan A (immunohistochemistry, ×200)

Answer

Ectopic extramammary Paget disease

Histopathological examination showed hyperkeratosis, acanthosis and atypical large pale cells with abundant vacuolated cytoplasm in the epidermis.

There was no dermal invasion [Figures 2a and b]. Tumor cells were positive for cytokeratin 7, but negative for cytokeratin 20, P63 and Melan A by immunohistochemistry [Figures 3a-d].

The lesion was totally excised with 2 mm negative margins and was lost to follow up.

Discussion

Extramammary Paget disease is a rare intraepidermal adenocarcinoma that primarily affects apocrine gland-bearing skin most commonly over the anogenital area. When the disease affects nonapocrine bearing skin, it is called ectopic extramammary Paget disease.[1] The pathophysiology of ectopic extramammary Paget disease is unknown. Sawada *et al*. reported the possibility that ectopic extramammary Paget disease originates potentially not only from the apocrine gland but also from the eccrine gland.[2] Clinically and histologically, there is no difference between ectopic extramammary Paget disease and conventional one.[2] Extramammary Paget disease is well known for its association with malignancy. Chanda reported the incidence of underlying adnexal carcinoma and concurrent internal malignancy associated with extramammary Paget disease as 24% and 12%, respectively.[3] The location of the underlying internal malignancy appears to correlate with the site of extramammary Paget disease. For example, the perianal and perineal location is associated with adenocarcinoma of the digestive system and genitourinary malignancy, respectively.[4]

Till now, there have been 46 cases of ectopic extramammary Paget disease in literature, of which only two cases have been reported in association with internal malignancy. The first case is ectopic extramammary Paget disease of the lateral thigh arising in association with sweat gland carcinoma of the same area,[5] the second being ectopic extramammary Paget disease in the midline of the abdomen with underlying colon adenocarcinoma.[6]

Whole-body evaluation for malignancy should be performed. For evaluation of internal neoplasm, positron emission tomography-computed tomography, upper and lower endoscopy and laboratory tests were recommended. However, the patient refused further testing.[7]

Extramammary Paget disease is clinically similar to Bowen disease and superficial basal cell carcinoma.[8] Thus, clinicians should keep in mind the possibility of ectopic extramammary Paget disease when there is treatment resistance skin lesion with eczematous morphology on the apocrine gland poor regions.

The diagnosis of extramammary Paget disease, in this case, was supported by immunohistochemical findings. Pagetoid cells in pagetoid Bowen disease usually demonstrate immunohistochemical features of squamous cells, which are negative for CK7 and positive for P63.[9] In this case, positivity for CK7 but negativity for p63 and Melan A supported the diagnosis of extramammary Paget disease, ruling out Bowen disease and melanoma in situ. A negative result of CK20 immunohistochemical stain supported the diagnosis of primary extramammary Paget disease rather than secondary extramammary Paget disease from an internal malignancy.[9] This case has been reported as a rare case of ectopic extramammary Paget disease appearing on an unusual site.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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