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QUIZ SECTION

Asymmetrically Pigmented Patch on the Vulvo-perineal Area: A Quiz

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A 50-year-old Caucasian woman presented with a 4-month history of a vulvo-perineal lesion, occasionally accompanied by day-time pruritus, especially before menstruation. At its onset, the lesion was pink-to-white in colour, but recently the patient noted an alarming change to dark brown (Fig. 1). She was in otherwise good health and denied taking any medications. Her medical history was notable for the surgical excision of a pancreatic adenoma two years before. A gynaecological examination and colposcopy showed normal findings. PAP smear and HPV-DNA tests were negative.

Dermoscopic analysis revealed an equivocal pigmented lesion. An incisional biopsy showed atypical neoplastic cells, positive for periodic acid-Schiff and cytokeratin 7 (Fig. 2) and surrounded by numerous reactive dendritic HMB-45-positive melanocytes. No dermal invasion was observed.

What is your diagnosis? See page 382 for answer.

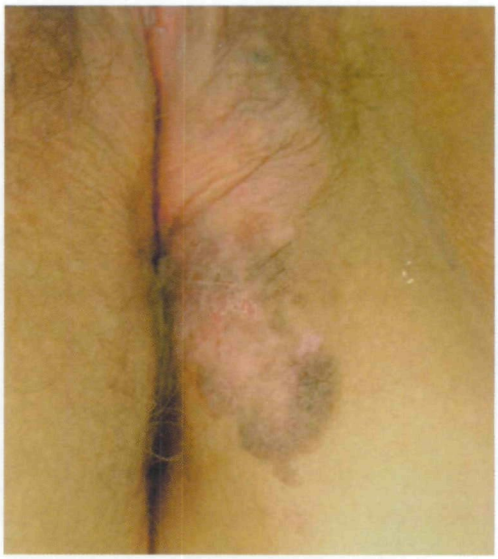

Fig. 1. Wide, asymmetrical brown lesion, partially dyschromic in the centre and with irregular borders, located on the left posterior vulvo-perineal region.

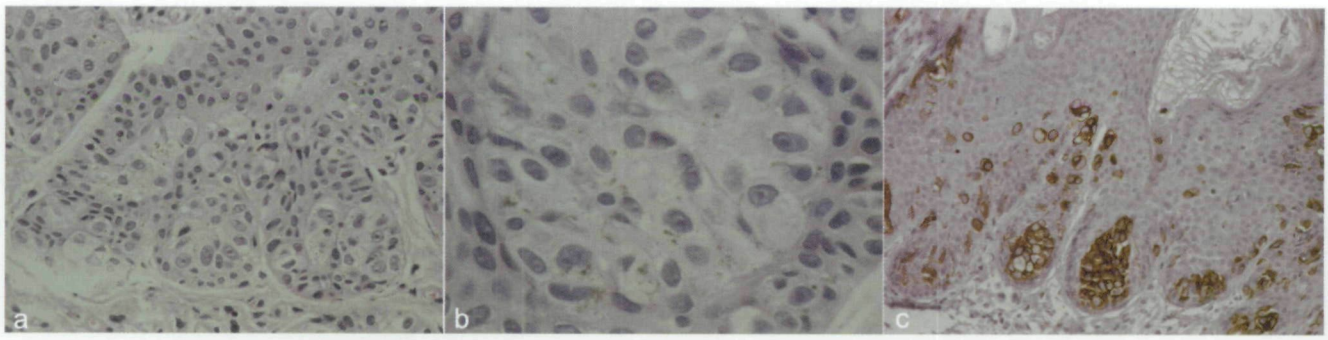

Fig. 2. a) Large, atypical cells, some clustered, in the epidermis. b) Neoplastic cells with abundant cytoplasm and vesicular nuclei, displaying focal gland differentiation (tubule formation). c) Immunohistochemical staining for cytokeratin 7.

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Tripe Palms, Digital Clubbing and Jaccoud's-type Arthropathy: A Quiz

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A 49-year-old man presented to the rheumatologist with a 12-month history of progressive deformity of the hands associated with thickening of the skin of the palms. He had a 60 pack/year history of cigarette smoking. Physical examination revealed Jaccoud's arthropathy (Fig. 1a), digital clubbing and hepatosplenomegaly. Cutaneous examination revealed focal hyperkeratosis of the palms, with a yellowish, verrucous appearance of the fingertips and accentuation of

ANSWERS TO QUIZ

Asymmetrically Pigmented Patch on the Vulvo-perineal Area: Comment

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Diagnosis: **Pigmented extramammary Paget's disease.**

The patient was subjected to blood chemistry and serum carcinoembryonic antigen tests, the results of which were all negative. Due to the previous surgical excision of a pancreatic adenoma, she also underwent abdominal and pelvic computer tomography and gastrointestinal endoscopy. Again the results were negative, as were those of a mammography. The patient underwent a partial left hemivulvectomy with deep resection to the superficial fascia and a Z-plastic procedure. Histological examination of a surgical specimen confirmed the diagnosis and excluded the presence of an underlying invasive neoplasm. At 28-months post-surgery the patient remains disease free.

Pigmented extramammary Paget's disease (PEMPD) is a very rare clinico-pathological variant of extramammary Paget's disease (EMPD) (1). In a small proportion of cases, EMPD is associated with underlying *in situ* or invasive neoplasia, or with distant internal organ neoplasms, most frequently gastrointestinal or urothelian (2, 3). Vulval localisation is rare, but typically occurs in postmenopausal Caucasian women. Clinically, it manifests as a uni- or multi-focal erythematous and/or eczematous lesion (2). Occasionally the lesion is pigmented (1). Dermoscopy is insufficient to produce a correct diagnosis, so differential diagnosis requires a histological biopsy to exclude malignant melanoma, heavily pigmented seborrhoeic keratosis and vulvar intraepithelial neoplasia. The histological typical features include reactive dendritic melanocytes in a process that is probably mediated by tumour-derived factors (4).

The standard treatment for primary vulval PEMPD does not differ from that for classical EMPD, i.e. surgical excision of the visible lesion with resection to the fascia and 2-cm gross margins (5). Although the prognosis following surgical resection is generally favourable, long-term monitoring of patients is recommended because recurrence is common and it is often necessary to repeat the surgical excision (6). For the preservation of the vulval anatomy, alternative non-surgical treatments have been used with conflicting results (7, 8).

The topical immunomodulatory cream imiquimod can be useful in cases in which surgery would be problematic because of the location and size of the tumour, as well as for recurrences (9). However, when we tested it in another EMPD-affected patient, control skin biopsies taken after the completion of therapy revealed residual tumour cells, although there was no clinical evidence of residual EMPD and pruritus was absent.

Photodynamic therapy based on the application of 5-aminolevulinic acid and subsequent irradiation with red light seems to have a place in treating recurrent forms of EMPD and in controlling symptoms (10), as well as in enhancing the efficacy of carbon dioxide laser ablation (11).

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Tripe Palms, Digital Clubbing and Jaccoud's-type Arthropathy: A Comment

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Diagnosis: **Pachydermatoglyphy in an apparently cancer-free smoker**

Tripe palms (pachydermatoglyphy) refers to an acquired palmoplantar keratoderma with enhancement of normal dermatoglyphics. It is frequently associated with neoplasia (1). Jaccoud's arthropathy is a chronic progressive painless deformity of the hands and feet, which is occasionally paraneoplastic (2).

The term "tripe palms" was initially coined by Clarke in 1977 (3). Approximately 100 cases of this rare paraneoplastic dermatosis have been described. Of these, 90% were associated with cancer. Pulmonary and gastric neoplasms are the most common associations (1, 4). In our patient an extensive search for an underlying malignancy was negative. After one