An Unusual Combination of Axillary Extramammary Paget’s Disease and Intradermal Naevus

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Case Report
A.B., born 1907, female, Caucasian.

History
An 84-year-old woman manifested for over a year a red, slowly expanding skin lesion in the left armpit. This lesion was discovered by chance and was asymptomatic. The patient was extraordinarily fit in view of her age.

Clinical Findings
The clinical examination showed a slightly elevated erythema-tous macula. The lesion was moderately infiltrated and measured 30×20 mm (fig. 1). There was no fixation with underlying structures in the armpit, and no deeper tumoral process could be observed. Further clinical examination revealed no other abnormalities.

Histology
A skin biopsy showed a tumoral cell population in the basal layers of the epidermis and a benign intradermal naevus in the underlying dermis (fig. 2). A mucin stain (PAS after amylase) showed the cell population only in the epidermis (fig. 3). The epithelial membrane antigen showed the same cell population (fig. 4). The cell population in the dermis was positively stained with protein S-100 but not the cell population in the epidermis (fig. 5). These findings excluded a melanoma in situ and confirmed the diagnosis of extramammary Paget’s disease in the armpit. The combination with an intradermal naevus seems to have been coincidental.
Fig. 1. Slightly elevated and erythematous macula in the left armpit.

Therapy
A broad local excision of the lesion was conducted under sedation and local anaesthesia. The procedure and postoperative course were uneventful.

Follow-Up
After 2 years, the follow-up is still satisfactory.

Laboratory Findings and Complementary Examinations A complete blood examination was normal. A dissemination examination consisting of a mammography, echography of the breasts, abdomen and pelvis, bone scintigraphy and thorax X-ray was also normal. It was concluded that it concerned a locally isolated focus of Paget’s disease in the armpit.

Discussion
Axillary extramammary Paget’s disease is a rare entity. It generally occurs in the genital regions [1]. In this case, it was a primary form of extramammary Paget’s disease since

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Fig. 2. Tumoral cell population in the basal layers of the epidermis and an intradermal naevus in the underlying dermis. HE. × 10.

Fig. 4. Epithelial membrane antigen (EMA) stains the epidermal cell population but not the dermal cell population. EMA. × 10.

Fig. 5. Protein S-100 stains the dermal cell population. Protein S-100. × 10.

No underlying internal malignancy could be found. This is the situation for the majority of such cases [1]. This pathology must be distinguished histologically very clearly from a Pagetoid form of Bowen’s disease and from an in situ melanoma [2]. Because of the presence of an intradermal naevus, an in situ melanoma had to be excluded with certainty. The complementary histochemical stains (PAS after amylase) and immunohistochemical stains (epithelial membrane antigen and protein S-100) were of exceptional importance for this purpose [3]. The intradermal naevus under the epidermal lesion, therefore, appeared to be purely coincidental.

Fig. 3. Mucin stain showing the cell population in the epidermis. PAS after amylase. ×25.

References