B. Sebaceous gland tumors

Sebaceous carcinoma

An orangey nodule occurs, most frequently on the Meibom glands in the palpebra sebaceous glands and less frequently in the skin (Fig. 22.17). Histopathologically, the tumor cell nest contains atypical clear sebaceous cells. In the autosomal dominantly inherited Muir-Torre syndrome, multiple benign or malignant sebaceous tumors occur, often accompanied by visceral malignancies.

C. Sweat gland tumors

1. Mammary Paget’s disease

Outline
- Infiltrative eczema-like erythema or erosion occurs in the nipples and at their periphery.
- It occurs most commonly in the opening of the lactiferous ducts of middle-aged women. It is a carcinoma in situ that derives from the lactiferous duct epithelia. It corresponds to breast cancer.
- A tumor does not form in most cases.
- It is not itchy, nor does it respond to steroids. Mammary Paget’s disease can be distinguished from eczema by these characteristics.
- The treatments are the same as those for breast cancer.

Clinical features

A plaque with clearly circumscribed erythema, infiltration and crusts appears on the nipple and areola (Fig. 22.18). The lesion is slightly firm and palpable, and usually unilateral. Middle-aged women are most frequently affected. Bilateral mammary Paget’s disease and mammary Paget’s disease in men are extremely rare. Mammary Paget’s disease accounts for 1% to 4% of all breast cancer cases. The symptoms progress gradually with each year. As they progress, a palpable tumor forms in the breast and metastasizes to a regional lymph node (mainly the axillary lymph node).

Pathogenesis

Mammary Paget’s disease is thought to originate from cancer in the excretory duct cells of the mammary glands (intraductal carcinoma) or carcinoma from epidermal keratinocytes.

Pathology

Large, clear Paget’s cells replace wall cells in the ducts and
glands. The Paget’s cells may also proliferate in cavities in the lactiferous ducts and mammary glands. Although the skin lesion does not clinically appear to be severe, Paget’s cells infiltrate lactiferous ducts and mammary glands more extensively than is clinically obvious. With further progression, Paget’s cells infiltrate into the dermis. Immunostaining is positive for CEA.

**Differential diagnosis**

Chronic breast eczema, tinea corporis, and basal cell carcinoma should be distinguished from mammary Paget’s disease. Intractable eczematous lesions on the breast that do not respond to topical agents should be suspected of being mammary Paget’s disease.

**Treatment**

The treatments are the same as those for breast cancer. Mastectomy and regional lymph node dissection are the basic treatments.

### 2. Extramammary Paget’s disease

**Outline**

- This is Paget’s disease on areas other than the breasts.

![Clinical images are available in hardcopy only.](a)

![Clinical images are available in hardcopy only.](b)

![Clinical images are available in hardcopy only.](c)

![Clinical images are available in hardcopy only.](d)

![Clinical images are available in hardcopy only.](e)

![Clinical images are available in hardcopy only.](f)

**Fig. 22.19 Extramammary Paget’s disease.**

The elderly are affected.
- Sharply margined eczematous erythema and erosion occur.
- It is thought to be intraepidermal cancer that originates from the apocrine glands. The genitalia, anal region and axillary fossae are most frequently involved.
- It occasionally destroys the basement membranes and progresses to invasive carcinoma.
- Extensive resection and lymph node dissection are necessary, if regional lymph node metastasis may occur.

**Clinical features**

Extramammary Paget’s disease occurs most commonly in the elderly. A bright red infiltrative plaque resembling mammary Paget’s disease appears (Fig. 22.19), most frequently on the genitalia, less frequently on the perianal region, perineum, axillary fossa or umbilical region. Itching is often present. The lesion gradually spreads, with melanin deposition at the periphery in some cases. Extramammary Paget’s disease occasionally destroys the basement membranes and develops a palpable small tumor in the lesion (Fig. 22.20). Regional lymph node metastasis occurs in advanced cases; the prognosis is poor.

**Pathogenesis**

Extramammary Paget’s disease is thought to originate from apocrine sweat gland cells.

**Pathology**

Large, bright, scattered or aggregated Paget’s cells are found in the epidermis and sweat ducts (Fig. 22.21).

**Differential diagnosis**

Eczema, candidiasis, genital tinea, Bowen’s disease, Hailey-Hailey disease and pemphigus vegetans are distinguished from extramammary Paget’s disease. Diagnosis can be made by pathological observation of Paget’s cells.

**Treatment**

The basic treatment is extensive surgical removal with a 1-to-3-cm margin including the peripheral normal skin.

3. Eccrine porocarcinoma

This is a malignant form of eccrine poroma (Chapter 21). A red plaque or nodule, often ulcerative, occurs, most frequently on the lower legs of the elderly (Fig. 22.22). In most cases, eccrine porocarcinoma is clinically observed as a tumor that is mix of eccrine poroma and eccrine porocarcinoma. It often metastasizes.

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**Clinical images are available in hardcopy only.**

Fig. 22.20 Invasive extramammary Paget’s disease that had been left untreated for a long period of time.

A flat lesion elevated gradually, forming infiltrative nodules. The lesion destroyed the basal membrane and infiltrated in the deep portions of the dermis. Metastasis to the regional lymph node was observed.

Fig. 22.21 Histopathology of extramammary Paget’s disease.

There are scattered Paget’s cells with large, clear cytoplasm.
Malignant Skin Tumors and Melanomas

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4. Microcystic adnexal carcinoma (MAC)

Synonym: Syringoid eccrine carcinoma

A firm, discoidal, intradermal nodule 1 cm to 3 cm in diameter occurs, most commonly around the mouth of persons of middle age and older. Some think it to be a sclerosing eccrine porocarcinoma or a cancer derived from a hair follicle or apocrine sweat gland. After extensive resection, the site should be examined pathologically for any remaining lesions because of the high frequency of local recurrence. For this reason, Moh’s microsurgery is also effective. Distant metastasis rarely occurs.

5. Mucinous carcinoma of the skin

A nodule of 2 cm to 3 cm in diameter occurs, frequently on the eccrine secretory part of the face or scalp. The tumor is covered by abundant mucin. The nuclei of tumor cells show slight atypism. Metastatic carcinoma of the skin with mucus production should be distinguished from mucinous carcinoma of the skin.

D. Nervous system tumors

Merkel cell carcinoma

Outline

• It is a skin cancer that originates from Merkel cells of the epidermis. These cells are thought to be tactile receptor cells.
• A highly malignant, domed red tumor forms on the face, head, neck or extremities of the elderly.
• Extensive resection, irradiation and chemotherapy are the main treatments.

Clinical features

A firm, domed nodule varying in color from light pink to purplish red and with a diameter of 1 cm to 3 cm occurs, most frequently on the face of the elderly (Fig. 22.23).

Pathology

Deep-staining small cells arrange densely in a palisading pattern, resembling the tumor cells of small-cell lung cancer (Fig. 22.24). Merkel cell carcinoma is characterized by dense-core granules that resemble Merkel cells (Fig. 22.25). Immunohistochemically, neuron specific enolase (NSE) and cytokeratin 20 are positive in many cases.