10. **Hidradenoma papilliferum**

A small, dome-shaped tumor occurs, often accompanied by erosion and bleeding. It most frequently appears on the female genitalia. The tumor resembles granulation tissue. Pathologically, there is dense papillary proliferation of glandular epithelial cells of apocrine-type secretion. Hidradenoma papilliferum is the typical type of apocrine neoplasm.

11. **Syringocystadenoma papilliferum**

An erosive rose-pink-surfaced verrucous nodule occurs, most commonly on the scalp or face of infants (Fig. 21.20). It is an apocrine organic hamartoma and often occurs secondarily after sebaceous nevus. Histopathologically, there is a double-layered luminal structure with long cylindrical cells on the inner side, cubical cells on the outer side, and marked plasmacytic interstitial infiltration in the nodule (Fig. 21.21). Basal cell carcinoma occurs secondarily in 10% of cases.

12. **Tubular apocrine adenoma**

A nodule of 1 cm to 2 cm in diameter and normal skin color or brown occurs, usually on the scalp. Histopathologically, proliferation of small, multiple cyst-like lumens is seen.

13. **Erosive adenomatosis of the nipple**

A benign tumor occurs in the nipple, often accompanied by erosion and exudation. Differentiation from mammary Paget’s disease and breast cancer is necessary. A dense concave structure and a luminal structure are histopathologically observed. Erosive adenomatosis of the nipple is a benign tumor that differentiates into the apocrine sweat glands. The only treatment is total excision; unless it is complete, there is recurrence.

E. **Cysts**

1. **Epidermal cyst**

Clinical features

A dome-shaped, intradermal or subcutaneous tumor with a diameter of 1 cm to 2 cm (or more than 10 cm in rare cases) occurs, most frequently on the head or neck, upper trunk, or lumbar region (Fig. 21.22). The tumor adheres to the skin surface;
however, the sides and bottom of the tumor mass do not firmly adhere to the peripheral tissue. It tends to occur on haired sites. Cysts are elastic but firm, with a surface color of normal skin or light blue and a black punctate opening at the center. They are asymptomatic. When pressed after excision, the cyst exudes a putrid-smelling, white, gruel-like discharge. Most cases clinically diagnosed as “atheroma” are epidermal cyst. Reddening, swelling and tenderness may be caused by secondary infection and rupture of the cyst walls (inflammatory epidermal cyst).

**Pathogenesis**

The epidermis or infundibulum-derived epithelial components invaginate into the dermis and proliferate to form a cyst that contains keratinous substances. Invagination of the epidermis or epithelial components into dermis that is caused by injury or by infection of HPV-57 or HPV-60 is thought to be associated with epidermal cysts in some cases, in which cases the cysts occur in the palms and soles (Table 23.1).

**Pathology**

The wall of the cyst has the same structure as normal epidermis: basal layer, suprabasal cell layer and granular cell layer (Fig. 21.23). However, instead of the horny cell layer, there are gruel-like, layered keratinous contents. When the keratinous substance is released into the dermis by rupture of the cystic wall, immunological reaction against foreign substance occurs, and a foreign body granuloma containing multiple polynuclear giant cells may be produced.

**Treatment**

The cyst and its walls are excised.

## 2. Milium

**Clinical features**

A small, firm, white to yellowish-white papule of 1 mm to 2 mm in diameter occurs immediately below the epidermis (Fig. 21.24). White keratinous contents are discharged by incision. Primary milium occurs most frequently on the eyelids, followed by the cheeks, penis and labia. Plaques may form. The histological findings are nearly the same as those of epidermal cyst.

**Definition, Pathogenesis**

The pathogenesis of primary milium is thought to be keratotic cyst formation resulting from abnormality of embryonic epithelial buds. Secondary milium occurs after a blistering disease (e.g., dystrophic epidermolysis bullosa, epidermolysis bullosa acquisita), burn scarring or radiodermatitis. The skin appendages and epidermal cells are damaged by these diseases and proliferate in cyst-like shape under the epidermis.
A small incision using a scalpel, or a puncture with a hypodermic needle, is conducted to remove the spherical white substance.

3. Dermoid cyst

A dome-shaped subcutaneous cyst with a diameter of 1 cm to 4 cm appears, most frequently on the head. It is present at birth. It is often misdiagnosed as an epidermal cyst. Histopathologically, sebaceous glands and sweat glands are found in the cyst walls that are produced by the epidermis.

4. Trichilemmal cyst

The head is affected in about 90% of cases. A trichilemmal cyst pathologically resembles an epidermal cyst. Histopathologically, there are epithelial cells that consist of cyst walls, and the epithelial cells on the luminal side of the cyst wall keratinize without forming a granular cell layer (trichilemmal keratinization, Fig. 21.25).

5. Steatocystoma multiplex

A firm, dome-shaped tumor with a diameter of 1 mm to 5 mm in most cases and a color ranging from that of normal skin to light yellow or light blue occurs, frequently on the axillary fossae, upper chest or upper arm (Fig. 21.26). It is follicle-associated in some cases. Some cases are autosomal dominant; mutation in the keratin 17 gene is associated with the condition. Histopathologically, there are flattened sebaceous glands near or directly attached to the tumor. The cyst wall is composed of intricately multilayered epithelial components.

6. Eruptive vellus hair cyst

This is an asymptomatic follicular papule that occurs most frequently on the chest. The cyst is superficial, crusted and umbilicated. It may be accompanied by steatocystoma multiplex. The cyst wall may contain a sebaceous structure.

7. Pilonidal sinus

Synonyms: Pilonidal cyst, Pilonidal disease

Young men whose buttocks, particularly their sacral division, are hairy are most frequently affected. Ingrowth of hair leads to formation of a fistula. The fistula is surrounded by granulomatous tissue or squamous epithelia. Pilonidal sinus may also occur on the occipital division, eyelids, genitalia, axillary fossae, umbilical fossae or interdigital areas. Most cases with interdigital
involvement occur occupationally, such as in barbers and shearers. The affected site, including the scar tissue, should be completely excised (Fig. 21.27).

8. Branchial cyst

A branchial cyst is an epidermal-cyst-like cyst that occurs on the preauricular region and neck. As it is caused by branchial debris, mobility is not fully present at the bottom of the cyst. There is a palpable cordlike substance in the cyst. Excision should not be decided quickly. Branchial cysts caused by thyro-lingual debris are called thyroglossal duct cysts.

9. Median raphe cyst of the penis

A tumor of several millimeters in diameter occurs in the penile raphe of young men (Fig. 21.28). It occurs solitarily at the urethral openings in most cases. The cyst may reach several centimeters in diameter. Its wall is histopathologically composed of single- or several-layered cylindrical epithelia or cubical epithelia that resemble urethral transitional epithelia.

F. Neural tumors

1. Neurofibroma

A neurofibroma is thought to be a benign tumor that derives from peripheral nerve Schwann cells or from perineurial or endoneurial cells. The tumor is sharply margined, dome-shaped, soft and of normal skin color or light pink (Figs. 21.29 and 21.30). It lacks a covering membrane and contains myxoid stroma. The tumor slowly enlarges (Figs. 21.29 and 21.30). There are almost no symptoms; however, subcutaneous neurofibroma (nodular plexiform neurofibroma) is often accompanied by tenderness. In neurofibromatosis type 1 (NF1), neurofibromas occur multiply on the whole body. In NF5, localized areas, such as on the trunk, may be affected by mosaicism. Nearly all of the subcutaneous tumors caused by NF1 are nervous neurofibromas, and neurilemmomas are not usually found (also see Chapter 20).

2. Neurilemmoma

Synonym: Schwannoma

Clinical features

A neurilemmoma is a Schwann-cell-derived benign tumor that