21.4). Thinning of the epidermis accompanied by parakeratosis is present in the concave center of the lesion.

**Treatment**

Excision, electrical coagulation, cryotherapy, dermabrasion, and administration of retinoids are the main treatments. Porokeratosis is chronic and intractable.

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**B. Follicular tumors**

**1. Trichofolliculoma**

Small, smooth-surfaced, dome-shaped tumors or papules 5 mm to 10 mm in diameter occur, most commonly in the nasal region and its peripheries ([Fig. 21.5](#fig21.5)). Trichofolliculoma is characterized by small keratotic cavities with several immature woolly hairs at the center. The pathogenesis is unknown. Trichofolliculoma is considered a benign tumor in which the entire follicle – including the inner root sheath, outer root sheath, and dermal hair papilla – differentiates.

**2. Trichoadenoma**

A firm, solitary, elastic nodule 1.5 cm or less in diameter appears, most frequently on the face. It is thought to be a tumor whose morphological differentiation falls between that of trichofolliculoma and that of trichoepithelioma. The border between the normal dermis and trichoadenoma is clear. There are multiple keratin-containing cyst(s) and solid masses of cells in the dermis.

**3. Solitary trichoepithelioma**

This is a benign tumor derived from hair germs that differentiate into various hair components, such as hair follicles, outer root sheaths, and hairs. Small, firm but elastic tumors of 2 mm to 5 mm in diameter and normal skin color occur around the nose, eyebrows, upper lip, and chin. It is nonhereditary. Solitary trichoepithelioma histopathologically consists of a small keratin-containing cyst(s) and basaloid cells, and there is proliferation of dermal stroma. It may be difficult to distinguish from basal cell carcinoma; however, in most cases of solitary trichoepithelioma,
there are well-differentiated keratinous cysts and formation, although incomplete, of hair follicles. Foreign body granuloma and calcium deposition may be present.

4. Trichoepithelioma papulosum multiplex

**Clinical features, Pathogenesis**

Multiple, dome-shaped, firm papules of several millimeters to 1 cm in diameter and normal skin color occur symmetrically on the midline facial region, scalp, nuchal and neck region, and trunk (Fig. 21.6). The onset is puberty, and the lesions gradually increase in number. Women are slightly more likely to be affected than men. It is autosomal dominant and may be familial. In recent years, abnormality in the cylindromatosis (CYLD1) gene has been identified as the cause.

**Pathology**

Trichoepithelioma papulosum multiplex is a tumor mass consisting of basaloid cells that resemble basal cell carcinoma cells. A keratinous cyst forms in the tumor.

**Differential diagnosis**

The papules produced on the midline facial region are similar to facial angiofibroma seen in tuberous disease (Chapter 20). In tuberous sclerosis, other symptoms such as leukoderma and shagreen patch are present.

**Treatment**

As malignant transformation is not present in trichoepithelioma papulosum multiplex, treatment and follow-up are unnecessary, except when there are cosmetic concerns.

5. Desmoplastic trichoepithelioma

Circular nodules or plaques of several millimeters to 1 cm in diameter and normal skin color or light yellow occur, most frequently on the cheeks, forehead and nasal region of relatively young adult women. The lesions are characterized by elevated edges and concave centers (Fig. 21.7). Miliary, papular lesions may occur at the periphery of the lesion. Histopathologically, cordlike proliferation of basaloid tumor cells, multiple keratinous cysts and hyalinized collagen fibrils are present. Differentiation from basal cell carcinoma may be difficult.

6. Trichoblastoma

A dome-shaped nodule occurs, most frequently on the face or scalp. It consists of fibrous interstitium and tumor cells that resemble follicular germinative cells. It may arise on sebaceous nevi. Differentiation from basal cell carcinoma may be difficult.
7. Pilomatrixoma

Synonyms: Calcifying epithelioma, Pilomatrixoma

Clinical features

A firm, intradermal or subcutaneous tumor 3 cm to 4 cm in diameter occurs on the face, neck or upper arms of infants, usually solitarily. The tumor surface is rough and the color is of normal skin or translucent bluish white. It is the firmness of bone (Fig. 21.8). Although it is usually asymptomatic, mild tenderness may be present. Myotonic dystrophy may induce multiple calcifying epithelioma. Malignant formation rarely occurs (pilomatrix carcinoma).

Pathogenesis

Calcifying epithelioma is a teratoma that originates from the hair follicle bulge. Some cases are caused by genetic abnormality in \( \beta \)-catenin.

Pathology

A sharply margined, irregularly shaped tumor mass appears in the lower dermal layer or subcutaneous tissue. The mass is not covered by a distinct membrane but is surrounded by fibrous connective tissue (Fig. 21.9). The tumor contains basaloid cells (originating from the hair matrix and staining basophilic) and shadow cells. Shadow cells are enucleated cells that stain eosinophilic. Foreign body granuloma and calcification are seen.

Treatment

The treatment is surgical removal.

8. Trichilemmoma

A tumor of 3 mm to 8 mm in diameter and normal skin color to light brown occurs, usually solitarily and most commonly on the face. Histopathologically, there are columnar cells arranged in a palisading pattern and a mass of clear cells that resemble outer root sheath cells. Malignant trichilemmoma occurs in rare cases.

9. Proliferating trichilemmal tumor

A subcutaneous nodule or tumor 1 cm to 10 cm in diameter occurs, most frequently on the scalp. It is pathologically similar to epidermal cyst and trichilemmal cyst (described later). Erosion and ulceration may be present on the surface. Trichilemmal keratinization is observed histopathologically. Overproliferation of cell components is also seen. Malignant proliferating trichilemmal tumor accompanied by atypism is pathologically differentiated from proliferating trichilemmal tumor.

Fig. 21.8 Calcifying epithelioma.
This is a subcutaneous nodule of 15 mm in diameter. It is accompanied by tenderness and light pink erythema.

Fig. 21.9 Histopathology of calcifying epithelioma.

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