

**Fig. 20.15-2 Ectopic Mongolian spot.**  
a: Forehead. b: Back. c: Lumbar region.



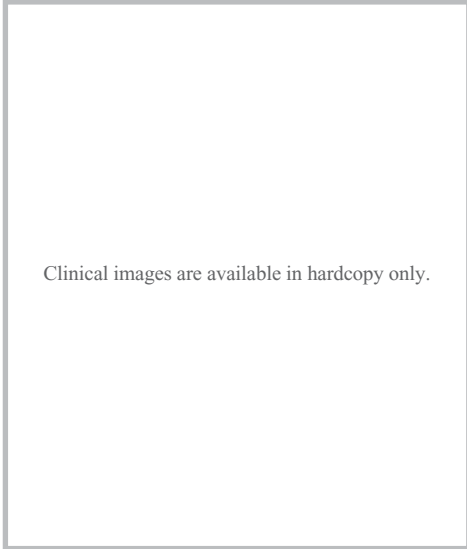
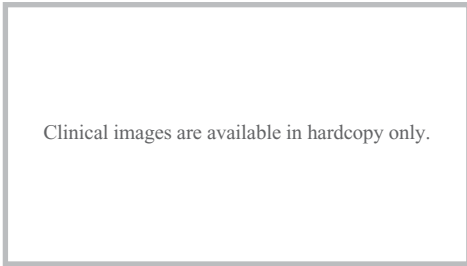
**Fig. 20.16 Laser treatment of aberrant Mongolian spot.**  
a: Mongolian spot on the shoulder and the right arm (pre-treatment). b: Mongolian spot after one session of alexandrite laser therapy. c: This portion (arrows) has improved significantly by alexandrite laser application.

## B. Epithelial nevi

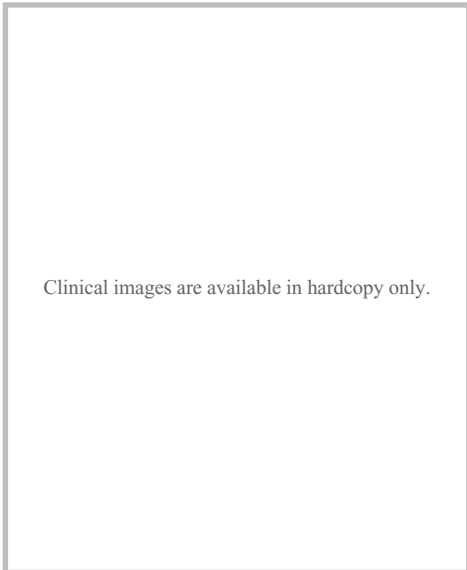
### 1. Epidermal nevus ★

#### Clinical features

Rough-surfaced, yellow to dark-brown papules or nodules are present at birth or in early childhood. They spread gradually, aggregate and form plaques of various sizes (**Fig. 20.17**). Although they may be localized, in most cases they are unilateral and arranged systematically along the Blaschko lines (**Fig. 1.2**). A generalized type spreads on the whole body.



**Fig. 20.17 Epidermal nevus.**



**Fig. 20.18 Inflammatory linear verrucous epidermal nevus.**

Although epidermal nevus tends to be asymptomatic, it may be accompanied by itching and eczematous changes. In inflammatory linear verrucous epidermal nevus (ILVEN), multiple, intensely itchy, light pink verrucous papules occur, most commonly on the lower legs of girls. They coalesce, become lichenified and arrange in linear pattern (**Fig. 20.18**).

Central nervous symptoms and skeletal abnormality are seen in rare cases; this is called epidermal nevus syndrome.

### Pathogenesis

Hyperplasia of epidermal keratinocytes results in localized or systematized verrucous nevus that enlarges gradually and becomes distinct.

### Pathology

Papilloma-like proliferation occurs in the epidermis. Granular degeneration also occurs in some cases. When ILVEN is accompanied by severer itching than usual, there is also inflammation, thickening of the epidermis, and parakeratosis.

### Treatment

Epidermal nevus can be left untreated, because changes and tumor formation rarely occur. Surgical excision, cryotherapy or laser therapy may be conducted for cosmetic improvement.

## 2. Nevus sebaceus ★

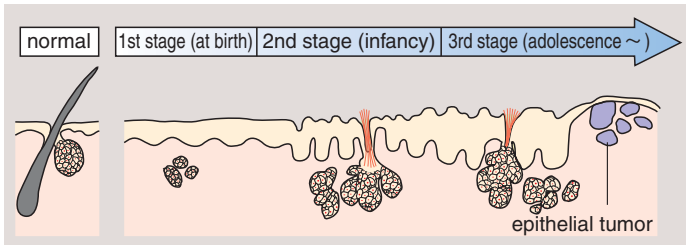
Synonym: Organoid nevus

### Outline

- It is caused by abnormal proliferation of various cells that originate in the epidermis, dermal appendages and connective tissue.
- It is present at birth. The scalp and face are most commonly affected. Nevus sebaceus on the scalp leads to alopecia.
- Removal is preferable, because malignant tumor such as basal cell carcinoma may develop.

### Clinical features

Nevus sebaceus occurs most frequently on the head and face (**Fig. 20.20**). The symptoms progress through three stages: first, at birth white to yellowish alopecia-areata-like plaques are present; second, with age the plaques elevate flatly, and gradually become verrucous and brownish; third, at puberty the lesions of the second stage worsen and additional epithelial tumors appear. Epithelial tumors such as dermal appendage tumors (e.g., syringocystadenoma papilliferum, trichoblastoma, outer root sheath tumor) and basal cell carcinomas may occur secondarily.



**Fig. 20.19 Histopathology of nevus sebaceus classified into three stages.**

The pathological and clinical features of the sebaceous glands gradually change with age.

**Pathology**

The histology differs for each of the three stages (**Fig. 20.21**). In the first stage, premature pilosebaceous tissue proliferates. In the second stage, there is maturation of the tissue, papilloma-like proliferation of the epidermis, ectopic proliferation of apocrine glands, and abnormality of dermal connective tissue. In the third stage, the histological changes of the second stage accelerate, with additional epithelial tumorous proliferation (**Fig. 20.19**).

**Treatment**

Surgical excision is conducted when secondary tumor is suspected or there are cosmetic concerns.

**3. Accessory mamma** ★

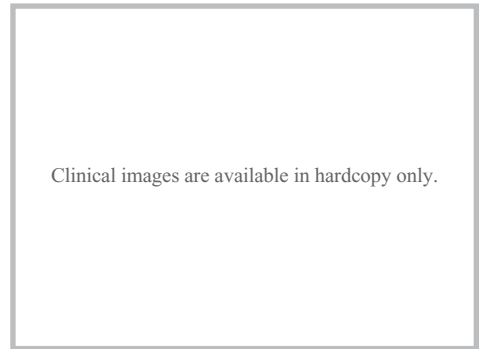
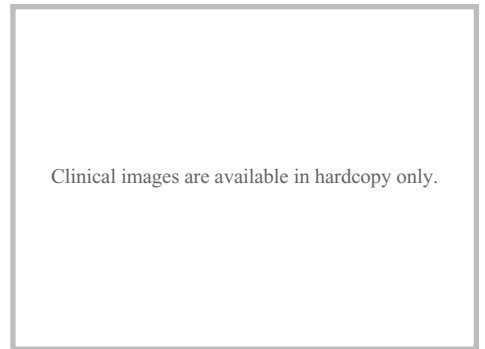
The primordia of the mammary glands, which exist along the line of the embryonic mammary ridge, normally disappear except for one pair in the chest. Accessory mamma is a condition in which more than one pair of primordia remains, usually on or around the axillary fossae. A brown patch or a palpable nodule of 1 cm to 2 cm in diameter appears on the axillary fossae or the anterior margin in most cases, accompanied by terminal hair. Swelling, pain and galactopoiesis may occur during pregnancy. Breast cancer may occur in rare cases.

**4. Nevus comedonicus**

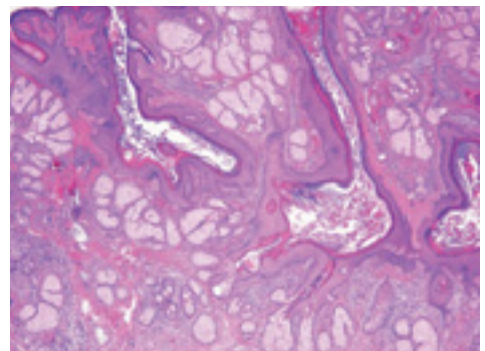
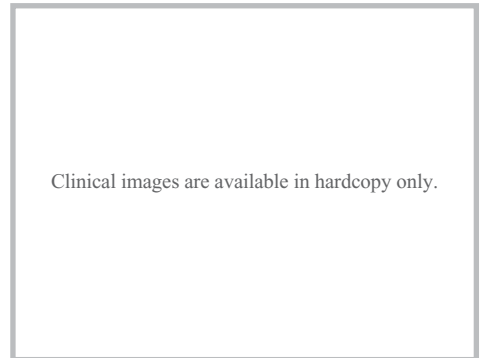
Dilated hair follicles with a black keratin plug aggregate or form a cord-like pattern (**Fig. 20.22**). They often occur between the time of birth and age 10. The face, neck, precordial region, abdomen and scalp are frequently affected.

**5. Hair follicle nevus**

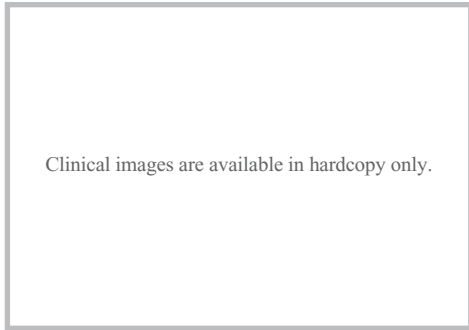
Dome-shaped or polyp-like papules or nodules of normal skin color are present at birth. The face is most frequently affected. Hair follicles in various stages of differentiation proliferate in all



**Fig. 20.20 Nevus sebaceus.**  
Nevus sebaceus and hair loss on the scalp.



**Fig. 20.21 Nevus sebaceus.**



**Fig. 20.22 Nevus comedonicus.**

dermal layers. The disorder may be accompanied by nevus sebaceus.

### 6. Eccrine nevus

Congenital localized hamartoma occurs in the eccrine sweat glands. A hyperhidrotic nodule and a plaque form. They occur between birth and infancy, most frequently on the extremities. When accompanied by angioma, it is called eccrine angiomatous hamartoma.

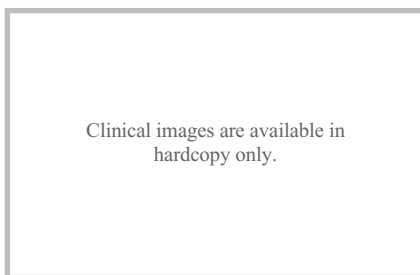
### 7. Apocrine nevus

Congenital localized hamartoma occurs in the apocrine sweat glands. The apocrine nevus is a papule or a small nodule, often accompanied by nevus sebaceous, which occurs on the scalp or axillary fossae.

## C. Mesenchymal-cell nevi



**Fig. 20.23 Connective tissue nevus.**



**Fig. 20.24 Smooth muscle hamartoma.**

### 1. Connective tissue nevus ★

Proliferation of dermal collagen fibers, elastic fibers or mucopolysaccharides results in eruptions of normal skin color that are called connective tissue nevi (**Fig. 20.23**). They may aggregate in patches.

### 2. Nevus lipomatosus cutaneus superficialis ★

Ectopic proliferation of fat cells in the dermis results in soft yellow nodules of several centimeters in diameter.

### 3. Nevus cartilagineus ★

Dome-shaped, cartilage-containing papules of normal skin color appear. Nevus cartilagineus in the ear region, called accessory ear, accompanies embryonic developmental failure of the branchial arch.

### 4. Smooth muscle hamartoma ★

This is a hamartoma in the arrector pili (**Fig. 20.24**). The lumbar and sacral regions are most commonly involved. The onset in most cases is within 6 months after birth. Slightly vaguely margined light brown patches appear. They may be hairy in some cases.

