Dermal melanocytes are produced in dermal melanocytic nevus, blue nevus, Mongolian spot and nevus of Ota. The distribution of cells varies according to the disease. Distributions of dermal melanocytes in blue nevus and other melanocytic nevi such as nevus of Ota and Mongolian spot are compared in Fig. 20.12.

1. Blue nevus

Outline

- A flat or slightly elevated blue nodule results from proliferation of melanocytes in the dermis (dermal melanocytes).
- It appears between the time of birth and infancy in most cases. The head, extremities and buttocks are most commonly involved.

Clinical features

A firm, blue or blackish small nodule of 1 cm or less in diameter appears. It may be flat or tumorous (Fig. 20.10). Blue nevus tends to appear singly and to progress gradually. The head, face, hands, legs, back and buttocks are most commonly affected.

Pathogenesis

Dermal melanocytes that have the ability to produce melanin and originate from neural crests become tumorous, causing blue nevus. The dermal melanocytes are filled with melanin granules that appear blue to brown on the skin.

Pathology, Diagnosis

There is tumorous proliferation of dermal melanocytes (Fig. 20.11). In nevocellular blue nevus, the dermal Schwann-cell-like cells lacking melanin production are seen. Blue nevus should be

Fig. 20.12 Classification of dermal melanocytic nevi by the distribution of melanocytes.
differentiated from malignant melanoma: blue nevus is characterized by its double-layered structure, and atypism and irregularity of size are not seen in the cell nuclei.

**Treatment, Prognosis**

The entire lesion is usually removed for cosmetic purposes. Careful clinical follow-up is necessary, because blue nevus may become malignant.

### 2. Nevus of Ota

**Synonym:** Nevus fuscocaeruleus ophthalmomaxillaris Ota

**Outline**

- Adolescent Asian women are most commonly affected. A light blue macule appears unilaterally on the skin at the first and second divisions of the trigeminal nerve. Melanosis of bulbar conjunctiva occurs.
- This nevus is caused by proliferation of dermal melanocytes.
- It is not malignant, nor does it heal spontaneously. Laser therapy is effective.

**Clinical features**

A light blue nevus occurs unilaterally on the skin over the first and second divisions of the trigeminal nerve (eyelids, zygomatic region, lateral forehead, cheek). The nevus is light blue and punctately dispersed with various other colors, including brown, red and dark blue (Figs. 20.13-1 and 20.13-2). It may be bilateral in some cases. Nevus of Ota with pigmentation in the sclera, iris and fundus is called oculodermal melanosis and is found in about half of cases. Pigmentation may also occur in the tympanic membranes, nasal membranes, pharynx and palate. A nevus with the same pigmentation as nevus of Ota but in the acromion and deltoid region is called nevus of Ito or nevus fuscocaeruleus acromiodeltoides Ito. It often causes great mental distress and cosmetic concern.

**Classification**

Nevus of Ota is classified into an early-onset type, in which pigmentation is present at birth and the color darkens as the patient grows, and a later-onset type, which occurs after puberty. Both types occur most frequently in Asian women and tend not to disappear spontaneously.

**Pathology**

Melanocytes are dispersed in the dermis. Pigmentation is present in the epidermal basal cell layer (Fig. 20.12).

**Treatment**

Laser therapy is extremely effective.
Acquired dermal melanocytosis used to be classified as a subtype of bilateral nevus of Ota; however, the two are now regarded as distinct diseases. Multiple, punctate, grayish-brown pigmentation of 1 mm to 3 mm in diameter occurs on both sides of the forehead and zygomatic region. The bulbar conjunctiva and palate are not involved (Fig. 20.14). Women between adolescence and middle age, particularly those of Japanese and Chinese descent, are most commonly affected. Histologically, there are dermal melanocytes in the upper dermal layer and pigmentation in the epidermal basal cell layer.

Mongolian spot occurs in the lumbosacral regions and buttocks of newborns. Nearly 100% of Asians, 80% to 90% of Africans (in whom the blue color tends to be invisible), and 1% to 20% of Caucasians are affected. It remains until adulthood in 3% to 4% of all cases. The blue hue intensifies until the age of 2 years and then gradually fades, disappearing by age 10. The Mongolian spot is differentiated from nevus of Ota by its lack of brownish tone. Mongolian spot on sites other than the lumbosacral regions and buttocks is called aberrant or ectopic Mongolian spot (Figs. 20.15-1 and 20.15-2), and the color tends not to fade spontaneously.

Mongolian spot is dermal melanosis in which dermal melanocytes from the embryonic period partially remain.

Treatment

Treatment is not necessary in most cases. Laser therapy may be conducted on large spots or ectopic Mongolian spots by the age of 2 or 3 years (Fig. 20.16).
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.

Clinical features

1. **Epidermal nevus**

   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.