“Nevus” is Latin for “maternal impression” or “birthmark.” It denotes a circumscribed, non-neoplastic skin or mucosal lesion. The term is qualified according to the cell or tissue of origin. Nevi may be caused by hereditary or embryologic factors and may appear at any time in life (Unna, 1894). They progress extremely slowly. Neurocutaneous syndrome includes nevi formed in the skin and nevoid lesions produced in the systemic organs that cause central nervous symptoms. Neurocutaneous syndrome is often categorized as a phacomatosis; however, that term has fallen out of use internationally in recent years. This chapter introduces the most common nevi. Angiomas are described in Chapter 21.

**Nevus**

**A. Melanocytic nevi** *(Fig. 20.1)*

- **a. Nevocellular nevus**
  
  Synonyms: Pigmented nevus, Nevus pigmentosus

  **Outline**
  
  - Nevocellular nevus is caused by proliferation of nevus cells. A small nevocellular nevus is commonly called a mole.
  - A hairy, giant, pigmented nevocellular nevus of 20 cm or more in diameter is called a giant congenital melanocytic nevus. It tends to progress to malignant melanoma.
  - Dermoscopic findings are important for diagnosis.
  - “Pigmented nevus” may be used as a synonym; however, non pigmented lesions are often seen.

  ![Fig. 20.1 Classification of melanocytic nevi.](image-url)
A nevocellular nevus is a flat-surfaced or verrucous macule or tumor that is brown, black, or sometimes normal skin color (Figs. 20.2-1 to 20.2-3). It may be accompanied by terminal hair. Nevocellular nevi are classified by size into three types. Small nevocellular nevus is commonly called mole or lentigo and varies in size from several millimeters to 1 cm in diameter. It is not present at birth but first appears between the ages of 3 and 4 and gradually increases in number and size to peak at puberty. After puberty, the color often fades and the nevus is replaced by fat tissue or fibrotic tissue. When the diameter exceeds 20 cm, the nevus is called a giant congenital melanocytic nevus.

Nevus cells are derived from neural crests and proliferate abnormally, resulting in blackish-brown pigmented macules. Melanocytes and Schwann cells are derived from neural crests; however, nevus cells do not differentiate into either of these (Fig. 20.3).

Nevocellular nevi are classified by location of proliferation into junctional nevus, intradermal nevus and compound nevus (Fig. 20.3).

Nevocellular nevus should be differentiated from freckles, lentigines, non-melanocytic lesions such as seborrheic keratosis, dermatofibroma, and most importantly early malignant melanoma (Chapter 22). Any pigmented lesion in adults that is growing or changing in any way should be examined carefully. When pigmentation spreads beyond the nail in the nail plate (Hutchinson’s sign), there is a high likelihood that a malignant melanoma is involved. Dermoscopic findings are important for diagnosis.

Even when the dermoscopic findings are benign, follow-up is necessary. Surgical removal is the basic treatment for cases in the palms and soles, which tend to have a high likelihood of malignancy, and in cases with a relatively large congenital nevocellular nevus. Laser therapy may be conducted if there are cosmetic concerns. Excision, ablation or skin grafting may be performed on a giant congenital melanocytic nevus. When it is too large for removal, long-term follow-up may be chosen to observe for any signs of malignant melanoma.
Nevus cells are localized in the dermo-epidermal junction. Junctional nevus is a compound of nevus cells that function similarly to melanocytes and resemble nevus cells morphologically. It is formed by large cubical cells that are able to produce melanin in great quantities.

This is a combination of junctional nevus and intradermal nevus. The nevi tend to be small and hyperpigmented.

Nevus cells are localized in relatively deep areas of the dermis (Fig. 20.4). Melanin production is markedly low in cells of intradermal nevus. The cells appear spindled, resembling Schwann cells.

Giant congenital melanocytic nevus is seen at birth, sometimes accompanied by black bristles (giant hairy nevus). It may be larger than 20 cm in diameter (Fig. 20.5). In several percent of cases, malignant melanoma may develop, sometimes accompanied by central nervous symptoms (neurocutaneous melatosis, described later). Surgical treatment is difficult.
2. Divided nevus

Divided nevi distribute predominantly on the upper and lower eyelids. With the eye closed, they appear to be a single lesion. The color is blackish-brown. They are found at birth in most cases. When occurring in the penis, it is a pigmented lesion that is separated by the coronal sulcus into the glans and the penis (Fig. 20.6).

3. Melanonychia due to nevocellular nevus

Black lines appear on the nail plate because of the presence of nevus cells in the nail bed (Fig. 20.7). Most cases are benign; however, there is a possibility of malignant melanoma.

4. Sutton nevus

It is nevus pigmentosus that is surrounded by leukodermas (Chapter 16).

5. Spitz nevus

Synonyms: Juvenile melanoma, Spindle and epithelioid cell nevus

**Outline**
- This specific subtype of nevocellular nevus frequently occurs in young adults.
- It appears suddenly on the face in most cases and enlarges quickly to about 1 cm in diameter. The periphery may become reddish.
- It may have clinically and histopathologically similar features to malignant melanoma; nevertheless, Spitz nevus is benign and may resolve spontaneously.
- Differential diagnosis from malignant melanoma is essential. Surgical removal is the first-line treatment.

**Clinical features**

A small, dome-shaped nodule, usually solitary and ranging in color from light pink to reddish-brown or black and ranging in size from several millimeters to 2 cm occurs, most commonly in children but also in adult men and women. It suddenly appears, mainly on the face but also elsewhere, and enlarges to about 1 cm in diameter (Fig. 20.8). Because it may be accompanied by dark brown pigmentation, Spitz nevus is sometimes difficult to differentiate from malignant melanoma. Spitz nevus is benign and does not enlarge beyond a certain size, nor does infiltration occur.

**Pathology, Diagnosis**

Spitz nevus is a compound nevus containing various cells,
including spindle cells, epithelioid-like cells and multinuclear cells. Dermal edema, telangiectasia, and inflammatory cell infiltration may occur. These findings resemble those of malignant melanoma; differentiation between Spitz nevus and malignant melanoma is often difficult. The basic structural pattern of nevocellular nevi is preserved in Spitz nevus: the lack of cellular atypism in the cells is important in distinguishing Spitz nevus from malignant melanoma. Homogenous nonstructural eosinophilic substances called Kamino bodies are found in the nevus cell nest in 60% of cases (Fig. 20.9). Dermoscopy shows sharply circumscribed pigmented lesions with a characteristic starburst pattern at the periphery.

**Treatment**

Excision is conducted. Spitz nevus does not aggravate; however, careful differentiation from malignant melanoma is necessary.

### 6. Dysplastic nevus

**Synonym:** Clark nevus

Dysplastic nevus occurs around puberty. A slightly elevated, flat-topped patch or a pigmented nevus larger than 6 mm in diameter occurs. A light brown or black, sometimes light pink, lesion with roughly margined pigmentation forms. Dysplastic nevus is basically a benign nevocellular nevus. When multiple familial atypical nevi and malignant melanoma occur it is called dysplastic nevus syndrome. Dysplastic nevus syndrome is autosomal dominantly inherited, and it frequently develops into malignant melanoma. Most patients are Caucasians, and there are few Asian cases. Histopathologically, many cases show compound nevi. Atypism is not usually found in the cells. Dermoscopic differentiation from superficial spreading melanoma is necessary.

### 7. Spotted grouped nevus

Small blackish-brown pigmented macules or nodules densely aggregate in brown to light-brown patches. This nevocellular nevus occurs frequently on the trunk and less frequently at other sites.

### 8. Balloon-cell nevus

This nevus consists of balloon cells that contain clear, large cytoplasm. The cells are thought to be degenerated nevus cells. Nevertheless, balloon-cell nevus cannot be distinguished clinically from ordinary nevocellular nevus.

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**Lentigo**

Most cases of the pigmentation that is commonly called lentigo are small nevocellular nevus. Lentigo simplex, a dermatological term, is a flat blackish-brown lesion of several millimeters in diameter caused by localized proliferation of epidermal melanocytes in which the nevus cells do not increase in number over time. Lentigo simplex is not usually present at birth, but it may appear around 3 years of age. It accompanies systemic diseases (neurocutaneous syndrome) including Peutz-Jeghers syndrome, Cronkhite-Canada syndrome, and LEOPARD syndrome.