D. Other nevi accompanied by skin pigmentation

1. Nevus spilus

Synonym: Speckled and lentiginous nevus

Nevus spilus is a sharply marginated brown macule that can be of various sizes and shapes (Table 20.1). There is a congenital type and a late-onset type whose onset is infancy or later. A late-onset nevus spilus called Becker’s nevus occurs most commonly on the chest and scapular region of adolescent boys (Section 2, below). Melanocytic nevocellular nevus cells do not proliferate. Enhanced localized melanin pigmentation in the epidermal basal layer is the main finding. When multiple nevus spilus occurs in infancy, there is the possibility of café-au-lait spots of neurofibromatosis type 1 and McCune-Albright syndrome. Laser therapy and concealing cosmetics are useful.

2. Becker’s nevus

Synonym: Nevus spilus tardivus

This is histopathologically similar to nevus spilus. Irregularly shaped, patchy, light brown pigmentation first appears. It coalesces with newly produced macules at the periphery and enlarges to a diameter of several centimeters to 20 cm (Fig. 20.25). Hypertrichosis is present in about half of all cases. Proliferation of smooth muscle fibers is found in the dermis: Some regard Becker’s nevus as a type of smooth muscle hamartoma. Q-switched laser therapy is more effective on Becker’s nevus than on nevus spilus.

3. Nevus depigmentosus

Leukoderma of various sizes appear. They become more distinct several months to 1 year after birth (Chapter 16).

Table 20.1 Comparison between congenital nevus spilus and nevus spilus tardivus.

<table>
<thead>
<tr>
<th>Age of onset</th>
<th>Present at birth</th>
<th>Shortly before and after adolescence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Commonly affected sites</td>
<td>Whole body</td>
<td>Shoulders, lumbar area</td>
</tr>
<tr>
<td>Complications</td>
<td>Neurofibromatosis in some cases</td>
<td>None</td>
</tr>
<tr>
<td>Hypertrichosis</td>
<td>No</td>
<td>Yes, in some cases</td>
</tr>
<tr>
<td>Color tone</td>
<td>Light brown to blackish-brown</td>
<td>Darker than that of congenital nevus spilus</td>
</tr>
<tr>
<td>Size of eruption</td>
<td>Various</td>
<td>Usually large</td>
</tr>
<tr>
<td>Histopathological findings</td>
<td>Elevated levels of melanin in basal keratinocytes</td>
<td>Increased melanin in basal keratinocytes, possibly changes in the epidermis and increased arrector pili muscle</td>
</tr>
</tbody>
</table>
A sharply circumscribed white patch occurs, often on the upper chest, when the skin is flushed by bathing or rubbing. Known to be capillary dysfunction (catecholamine sensitivity), it may accompany neurofibromatosis type 1 (NF1) or nodular sclerosis.

**Synonym:** Von Recklinghausen disease

It is a neurocutaneous syndrome caused by proliferation of cells that originate from neural crests. Neurofibroma, pigmentation and nervous tumor in multiple organs are the main symptoms.

It is autosomal dominantly inherited, characterized by multiple pigmentation (café-au-lait spots) at birth and soft tumors (neurofibromas) after infancy.

Surgical removal and laser therapy are conducted.

**Clinical features**

1. **Café-au-lait spot**
   This is a flat, oval eruption of the color of coffee with cream or darker brown. It varies in size and appears on the trunk and extremities. A café-au-lait spot with a diameter of about 10 cm is called a large Recklinghausen spot; one with a diameter of 1 cm or less is called a small Recklinghausen spot. Individual eruptions cannot be distinguished from nevus spilus. A café-au-lait spot on the axillary fossae, called axillary freckling, is thought to be a specific symptom of NF1. The pigmentation of the spot is solid (**Figs. 20.27-1** and **20.27-2**). Café-au-lait spots are present in 70% of all newborns. After infancy, the number of spots does not increase.

2. **Neurofibroma**
   A neurofibroma is a soft tumor of normal skin color or light brownish-pink of various sizes (**Figs. 20.28-1** and **20.28-2**). It may be produced on any site of skin. It may be elevated and dome-shaped or papillary, or flat and palpable. Valvular or hanging...