ages of 10 and 30 (Figs. 21.66-1 and 21.66-2). The big toes are most frequently involved. Intense pain is present. Subungual exostosis is differentiated from glomus tumor. X-ray is useful. Excision is the main treatment.

Synonyms: Lymphadenosis benigna cutis, Pseudolymphoma

A lymphoid follicle structure forms as the result of an insect sting, external injury, sunlight or Lyme disease, leading to a dark red, dome-shaped tumor 1 cm to 2 cm in diameter, usually on the face (Fig. 21.67). The lesion is elastic and smooth-surfaced. Ulceration does not occur. The lesion appears solitarily in most cases and disappears spontaneously several months after onset. Differentiation from cutaneous B-cell lymphoma is important; follicle formation is the main characteristic of lymphocytoma cutis, and atypism is not found in lymphocytes. Lymphocytoma cutis has a good prognosis, although it progresses to lymphoma in rare cases.

1. Lymphocytoma cutis

Syonyms: Lymphadenosis benigna cutis, Pseudolymphoma

An asymptomatic, infiltrative plaque ranging in color from light pink to reddish brown occurs, most frequently on the face. Although it disappears spontaneously, it may recur. Dense lymphocytic infiltration is found in the dermis, especially in the peripheral appendages. Lymphocytic infiltration of the skin is differentiated from discoid lupus erythematosus and lymphoma.
3. Kimura’s disease

Kimura’s disease, whose cause is unknown, occurs commonly on the face of pubertal men. Cutaneous lymphoreticular tissue proliferates reactively to cause the disease. Solitary or multiple, flat or dome-shaped, soft, elastic, partially nodular, and subcutaneous or intradermal tumors of 5 cm to 10 cm in diameter appear. The surface of the lesion is brownish, and itching may be present (Fig. 21.68). Subcutaneous lymphatic follicle formation and eosinophilic infiltration are observed histopathologically. Kimura’s disease is characterized by the marked increase of eosinophils in the peripheral blood and bone marrow, and elevated IgE level. It may be accompanied by atopic dermatitis and pruritus. Local steroid injection is effective. Differentiation between Kimura’s disease and angiolymphoid hyperplasia with eosinophilia (ALHE, see the next section) has been controversial.

4. Angiolymphoid hyperplasia with eosinophilia (ALHE)

A firm, bright or dark red nodule of several centimeters in diameter occurs, frequently on the peripheral auriculae, forehead or temporal area (Fig. 21.69). The skin lesion is vascular proliferation of epithelial cells that contain abundant cytoplasm. Dense infiltration of eosinophils and lymphocytes is often found in the peripheral blood vessels. Although angiolymphoid hyperplasia with eosinophilia (ALHE) has some similarities with Kimura’s disease, these two diseases are in different categories. The main treatment for ALHE is local injection of steroids; nevertheless, the disease is intractable. Dye laser therapy is effective in some cases.

5. Mastocytosis

Synonyms: Urticaria pigmentosa, Mastocytoma

**Outline**

- Mast cells proliferate and become tumorous.
- Urticaria is caused by rubbing (Darier’s sign).
- It occurs most frequently in infants, healing spontaneously by adulthood. When the onset is in adulthood, the disease is intractable.
- Urticarial attacks may recur in some cases.

**Clinical features**

The onset of mastocytosis is in the first year after birth, in most cases. The adult type, which is rare, has an onset of puberty or thereafter. In infant mastocytosis, multiple round or spindled brown patches or small nodules of 1 cm or less in diameter occur after recurrent urticaria on the face and trunk. A solitary nodule of several centimeters may occur in rare cases (Table 21.1).
When mechanical stimulation is given to sites with eruptions, histamine is released from the mast cells, leading to the formation of urticaria (Darier’s sign, Figs. 21.70 and 21.71). Urticaria may be caused on the whole body skin by bathing or rubbing with a towel, leading to systemic symptoms such as flushing, nausea, vomiting, diarrhea, stomachache, fever, cardiac palpitation, breathing difficulty and shock (urticarial attacks).

In adult mastocytosis, these symptoms first appear at puberty or thereafter, and the eruptions and systemic symptoms tend to be moderate. Darier’s sign is not significantly noticeable. In some cases, extremely itchy diffuse eruptions may occur. There is malignant formation in rare cases. Systemic mastocytosis is accompanied by lymph node enlargement, splenohepatomegaly, osteoporosis and osteosclerosis. Thrombocytopenic bleeding tendency is present. It may become leukemia (mast-cell leukemia).

**Classification, Pathogenesis**

Mast cells that become tumorous and proliferate in the skin or in the whole body are stimulated, leading to the release of histamine and heparin, which results in urticaria. Mastocytosis in which localized cutaneous lesions occur is called mastocytoma. When tumorous lesions spread to the bone marrow, gastrointestinal tract or spleen, it is called systemic mastocytosis. The pathogenesis is unknown.

**Pathology**

There is proliferation of polygonal mast cells of various sizes. In the upper dermal layer, there is abnormal proliferation of polygonal mast cells of various sizes that stain metachromatically in toluidine blue (Fig. 21.72). It is classified by the proliferative pattern into Unna mastocytosis and Róna mastocytosis. In the former, multiple proliferative foci form map-like shapes resembling islands. In the latter, a few dispersed perivascular foci form.

**Table 21.1 Classification of mastocytosis**

<table>
<thead>
<tr>
<th>Condition</th>
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<tbody>
<tr>
<td>Urticaria pigmentosa</td>
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<tr>
<td>(Solitary) mastocytoma</td>
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<tr>
<td>Diffuse cutaneous mastocytosis</td>
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<tr>
<td>Telangiectasia macularis eruptiva perstans</td>
</tr>
</tbody>
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Clinical images are available in hardcopy only.

**Fig. 21.70** Mastocytosis.

**Fig. 21.71** Mastocytosis.

a: Solitary mastocytosis in an infant. b: Darier’s sign: urticaria is artificially caused by mechanical stimulation. c: Mastocytosis with blistering.
In pigmented areas of the skin lesion, the concentration of melanin increases in the epidermal suprabasal cell layer and basal layer in pigmented areas of the skin lesion.

**Treatment, Prognosis**

Any stimulation that may induce release of histamine, such as bathing or rubbing the skin, should be avoided. Treatment for urticarial attacks is that same as for general urticaria (administration of histamine). Infant mastocytosis heals spontaneously in several years to over a dozen years. It does not need treatment, as long as there are few eruptions and no severe attacks. Adult mastocytosis does not heal spontaneously and is intractable.

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**Fig. 21.72 Histopathology of mastocytosis (Unna mastocytosis).**