These disorders are caused by deposition or accumulation of amorphous glycoproteins, called amyloids, in the tissue or intercellular spaces. Amyloids consist of various precursor substances whose compositions differ according to the type of disease. Localized cutaneous amyloidosis occurs only in the skin; systemic amyloidosis affects systemic internal organs (Table 17.1).

Amyloids deposit and accumulate in the tissue and intercellular spaces, inducing dysfunction in the whole body or specific organs. Amyloids are glycoproteins that have a fibrous structure. They are not seen in normal metabolism. They consist of various precursors, such as immunoglobulin L-chains, abnormal prealbumin, and serum proteins. In each disease, the amyloids have a different composition. Cutaneous amyloidoses are classified as shown in Table 17.1.

## Outline
- These disorders are caused by deposition or accumulation of amorphous glycoproteins, called amyloids, in the tissue or intercellular spaces.
- Amyloids consist of various precursor substances whose compositions differ according to the type of disease.
- Localized cutaneous amyloidosis occurs only in the skin; systemic amyloidosis affects systemic internal organs (Table 17.1).

## Classification, Pathogenesis
Amyloids deposit and accumulate in the tissue and intercellular spaces, inducing dysfunction in the whole body or specific organs. Amyloids are glycoproteins that have a fibrous structure. They are not seen in normal metabolism. They consist of various precursors, such as immunoglobulin L-chains, abnormal prealbumin, and serum proteins. In each disease, the amyloids have a different composition. Cutaneous amyloidoses are classified as shown in Table 17.1.

## Pathology, Laboratory findings
Amyloids stain light red with PAS and orange-red with Congo red, and appear green to fluorescent yellow in polarizing microscopy. Localized cutaneous amyloidosis does not readily stain with Congo red; methyl violet (purple), thioflavine (fluorescent yellow), and dylon staining (reddish brown) are used instead (Fig. 17.1). Detection of Bence Jones proteins in the urine and detection of M proteins by electrophoresis of the serum has diagnostic value for systemic amyloidosis.

## Treatment, Prognosis
Topical steroids are effective against localized cutaneous amyloidosis, which has a good prognosis. Systemic amyloidosis accompanied by myeloma has a poor prognosis. Most patients die from renal dysfunction or heart failure within several years after onset.
a. Localized cutaneous amyloidoses

1. **Lichen amyloidosis**

   Lichen amyloidosis frequently occurs on the extensor surfaces of the lower legs, and on the forearms and back. Multiple, flat-surfaced, smooth, light-brown papules appear and may aggregate (Fig. 17.2). Intense itching is present in most cases. Histopathologically, the horny cell layer and epidermis thicken diffusely, melanin granules increase in the basal layer, and amyloid accumulates in the dermal papillae. Topical steroids and oral histamines are effective.

2. **Macular amyloidosis**

   Punctuate or reticular pigmentation occurs, most commonly on the scapular region and back of middle-aged women. Chronic rubbing of the skin with nylon towels causes ripple pigmentation (friction melanosis; Chapter 16). Amyloids may deposit in the skin. Friction melanosis is thought to be strongly associated with macular amyloidosis.

3. **Nodular localized cutaneous amyloidosis**

   **Synonym:** Amyloidosis cutis nodularis atrophicans

   It most frequently occurs in women middle-aged and older. Tan nodules appear on the lower abdomen. Atrophy of the dermis occurs. Diffuse amyloid deposition is found directly below the epidermis and fat tissue (Fig. 17.3).

4. **Poikiloderma-like cutaneous amyloidosis**

   Amyloid deposition is found in skin with poikiloderma.

5. **Anosacral cutaneous amyloidosis**

   This occurs in the anus and sacral region of the elderly. It is clinically characterized by pigmentation accompanied by hyperkeratosis. Amyloid deposition is seen pathologically.

6. **Secondary localized cutaneous amyloidosis**

   Deposition of amyloids is observed histologically in the dermal papilla in association with various skin disorders. Lesions including those of seborrheic keratosis, actinic keratosis, basal cell carcinoma, Bowen’s disease, cylindroma, calcified epithelioma, nevus sebaceus, verruca vulgaris, malignant lentigo, mycosis fungoides, psoriasis, discoid lupus erythematosus, lichen simplex chronicus, and solar dermatitis occur.