AL amyloidosis occurs frequently in men and women in their 60s. The cause is known to be plasma cell dyscrasia (such as multiple myeloma), in which abnormal immunoglobulin-related amyloid light chain (AL) was produced. Shiny yellowish-white papules occur, most frequently in the eyelids and often accompanied by purpura. Amyloid deposition is found among the collagen fibers in the epidermis and outer membranes of the blood vessels at the sites around the eruptions. Such deposition is also seen in the systemic organs, such as the gastrointestinal tract, cardiac muscles and skeletal muscles, where it causes various symptoms (Fig 17.4a). Lesions develop in the oral cavity and laryngeal mucosa, resulting in macroglossia and hoarseness. Generalized scleroderma-like stiffness in the fingers and nail deformity are present (Figs. 17.4b and 17.4c). Bence Jones proteins are excreted in the urine in some cases (a finding of myeloma). Treatments are mainly made for plasma cell dyscrasia. It has a poor prognosis as a result of the complication of cardiac insufficiency; most patients die within 2 years after onset.

The precursor protein of AA amyloidosis is serum amyloid A (AA) protein. The disorder is caused secondarily by a chronic inflammatory disease or an infectious disease, such as rheumatoid arthritis, tuberculosis or leprosy. Skin lesions rarely form. This disorder is autosomal dominantly inherited. Amyloid deposition in various organs, including the nerves, stomach and heart, causes dysfunction.

Hemodialysis-related amyloidosis

This occurs in those who have a prolonged hemodialysis. \( \beta_2 \)-microglobulin, which is not readily removed by hemodialysis, deposits as amyloids. The intercarpal synovial membranes, joints, heart, blood vessels, digestive tract and kidneys are affected. Erythema, papules, purpura and subcutaneous nodules are the main cutaneous symptoms.