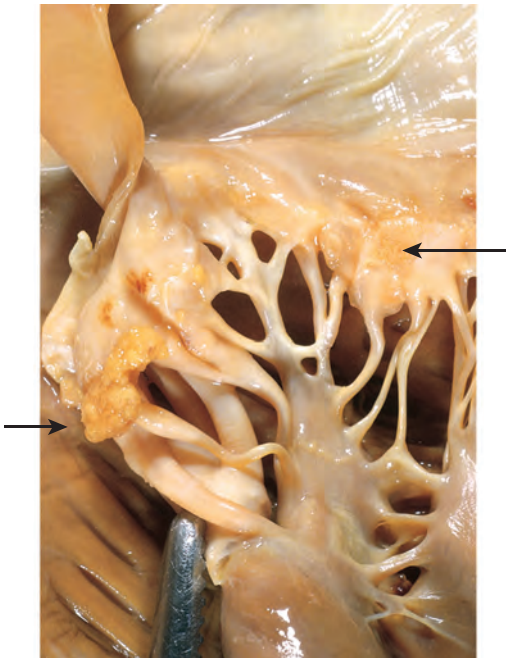

Figure 6-28 Libman-Sacks endocarditis of the mitral valve in lupus erythematosus. The vegetations attached to the margin of the thickened valve leaflet are indicated by *arrows*. (Courtesy Dr. Fred Schoen, Department of Pathology, Brigham and Women's Hospital, Boston, Mass.)

vegetations in infective endocarditis are considerably larger, and those in rheumatic heart disease (Chapter 12) are smaller and confined to the lines of closure of the valve leaflets.

An increasing number of patients have clinical evidence of coronary artery disease (angina, myocardial infarction) owing to coronary atherosclerosis. This complication is particularly notable in young patients with long-standing disease, and especially prevalent in those who have been treated with corticosteroids. The pathogenesis of accelerated coronary atherosclerosis is unclear but is probably multifactorial. Risk factors for atherosclerosis, including hypertension, obesity, and hyperlipidemia, are more commonly present in SLE patients than in the population at large. In addition, immune complexes and antiphospholipid antibodies may cause endothelial damage and promote atherosclerosis.

Spleen. Splenomegaly, capsular thickening, and follicular hyperplasia are common features. Central penicilliary arteries may show concentric intimal and smooth muscle cell hyperplasia, producing so-called onion-skin lesions.

Lungs. In addition to pleuritis and pleural effusions, which are present in almost 50% of patients, in some cases, there is chronic interstitial fibrosis and secondary pulmonary hypertension. None of these changes is specific for SLE.

Other Organs and Tissues. LE, or hematoxylin, bodies in the bone marrow or other organs are strongly indicative of SLE. Lymph nodes may be enlarged with hyperplastic follicles or even demonstrate necrotizing lymphadenitis.

Clinical Features. SLE is a multisystem disease that is highly variable in its clinical presentation, and its diagnosis relies on a constellation of clinical, serologic, and morphologic changes (Table 6-9). Typically, the patient is a young woman with some, but not necessarily all, of the following features: a butterfly rash over the face, fever, pain but no deformity in one or more peripheral joints (feet, ankles, knees, hips, fingers, wrists, elbows, shoulders), pleuritic chest pain, and photosensitivity. In many patients, however, the presentation of SLE is subtle and puzzling, taking forms such as a febrile illness of unknown origin, abnormal urinary findings, or joint disease masquerading as rheumatoid arthritis or rheumatic fever. "Generic" ANAs, detected by immunofluorescence assays, are found in virtually 100% of patients, but these are not specific for SLE. A variety of clinical findings may point toward renal involvement, including hematuria, red cell casts, proteinuria, and in some cases the classic nephrotic syndrome (Chapter 20). Laboratory evidence of some hematologic derangement is seen in virtually every case, but in some patients anemia or thrombocytopenia may be the presenting manifestation as well as the dominant clinical problem. In still others, mental aberrations, including psychosis or convulsions, or coronary artery disease may be prominent clinical problems. Patients with SLE are also prone to infections, presumably because of their underlying immune dysfunction and treatment with immunosuppressive drugs.

The course of the disease is variable and unpredictable. Rare acute cases result in death within weeks to months. More often, with appropriate therapy, the disease is characterized by flare-ups and remissions spanning a period of years or even decades. During acute flare-ups, increased formation of immune complexes results in complement activation, often leading to hypocomplementemia. Disease flares are usually treated with corticosteroids or other immunosuppressive drugs. Even without therapy, in some patients the disease may run an indolent course with relatively mild manifestations, such as skin changes and mild hematuria, for years. The outcome has improved significantly, and an approximately 90% 5-year and 80% 10-year survival can be expected. The most common causes of death are renal failure and intercurrent infections. Coronary artery disease is also becoming an important cause of death. Patients treated with steroids and immunosuppressive drugs incur the usual risks associated with such therapy.

As mentioned earlier, involvement of skin along with multisystem disease is fairly common in SLE. The following sections describe two syndromes in which the cutaneous involvement is the exclusive or most prominent feature.

Chronic Discoid Lupus Erythematosus. Chronic discoid lupus erythematosus is a disease in which the skin manifestations may mimic SLE, but systemic manifestations are rare. It is characterized by the presence of skin plaques showing varying degrees of edema, erythema, scaliness, follicular plugging, and skin atrophy surrounded by an elevated erythematous border. The face and scalp are usually affected, but widely disseminated lesions occasionally occur. The disease is usually confined to the skin, but 5% to 10% of patients with discoid lupus erythematosus develop multisystem manifestations after many years. Conversely, some patients with SLE may have prominent discoid lesions in the skin. Approximately 35% of patients show a positive test for generic ANAs, but antibodies