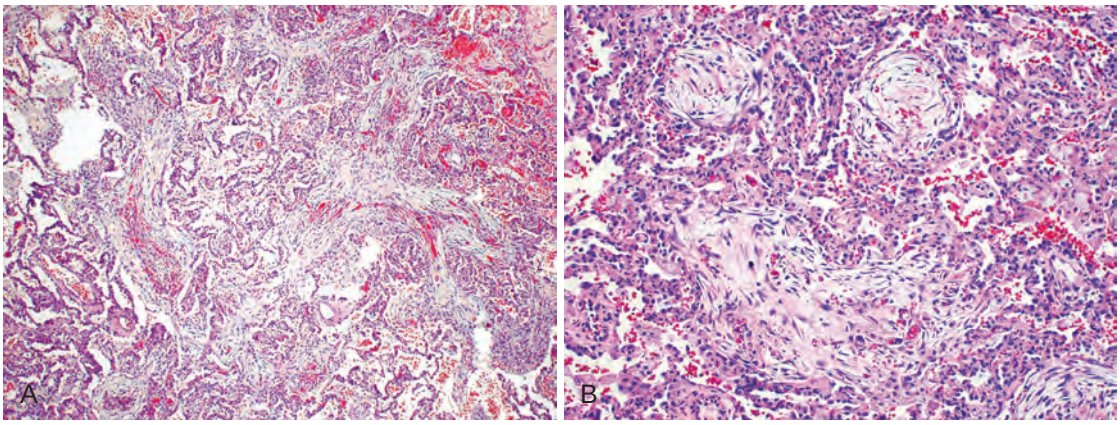

Figure 15-16 Cryptogenic organizing pneumonia. Some alveolar spaces are filled with balls of fibroblasts (Masson bodies), while the alveolar walls are relatively normal. **A,** Low power. **B,** High power.

Cryptogenic Organizing Pneumonia

Cryptogenic organizing pneumonia is synonymous with the popular term *bronchiolitis obliterans organizing pneumonia*; however, the former is now preferred, since it conveys the essential features of a clinicopathologic syndrome of unknown etiology and avoids confusion with airway diseases such as bronchiolitis obliterans. Patients present with cough and dyspnea and have patchy subpleural or peribronchial areas of airspace consolidation radiographically. Histologically, cryptogenic organizing pneumonia is characterized by the presence of polypoid plugs of loose organizing connective tissue (Masson bodies) within alveolar ducts, alveoli, and often bronchioles (Fig. 15-16). The connective tissue is all of the same age, and the underlying lung architecture is normal. There is no interstitial fibrosis or honeycomb lung. Some patients recover spontaneously, but most need treatment with oral steroids for 6 months or longer for complete recovery.

It is important to recognize that organizing pneumonia with intra-alveolar fibrosis is most often seen as a response to infections or inflammatory injury of the lungs. These include viral and bacterial pneumonia, inhaled toxins, drugs, connective tissue disease, and graft-versus-host disease in bone marrow transplant recipients. The prognosis for these patients is dependent on the underlying disorder.

Pulmonary Involvement in Autoimmune Diseases

Many autoimmune diseases (also referred to a connective tissue diseases because of their frequent association with arthritis), most notably systemic lupus erythematosus, rheumatoid arthritis, progressive systemic sclerosis (scleroderma), and dermatomyositis-polymyositis, can involve the lung at some point in their course. Pulmonary involvement can take different histologic patterns; nonspecific interstitial pneumonia, usual interstitial pneumonia, vascular sclerosis, organizing pneumonia, and bronchiolitis are the most common.

- *Rheumatoid arthritis*: pulmonary involvement may occur in 30% to 40% of patients as (1) chronic pleuritis, with or without effusion; (2) diffuse interstitial pneumonitis and fibrosis; (3) intrapulmonary rheumatoid nodules; (4) follicular bronchiolitis; or (5) pulmonary hypertension
- *Systemic sclerosis* (scleroderma): diffuse interstitial fibrosis (nonspecific interstitial pattern more common than usual interstitial pattern) and pleural involvement
- *Lupus erythematosus*: patchy, transient parenchymal infiltrates, or occasionally severe lupus pneumonitis, as well as pleurisy and pleural effusions.

Pulmonary involvement in these diseases has a variable prognosis that is determined by the extent and histologic pattern of involvement.

KEY CONCEPTS

Chronic Interstitial Lung Diseases

- Diffuse interstitial fibrosis of the lung gives rise to restrictive lung diseases characterized by reduced lung compliance and reduced forced vital capacity (FVC). The ratio of FEV_1 to FVC is normal.
- Idiopathic pulmonary fibrosis is prototypic of restrictive lung diseases. It is characterized by patchy interstitial fibrosis fibroblastic foci and formation of cystic spaces (honeycomb lung). This histologic pattern is known as usual interstitial pneumonia.
- The cause of idiopathic pulmonary fibrosis is unknown, but genetic analyses point to roles for senescence of alveolar epithelium (due to telomere shortening), cell stress related to protein misfolding, abnormal signaling in alveolar fibroblasts, and altered mucin production. The resulting injury to alveolar epithelial cells set in motion event that lead to increase local production of fibrogenic cytokines such as TGF-β
- The other diseases that cause diffuse interstitial fibrosis are heterogeneous poorly understood, but most have better prognoses that idiopathic pulmonary fibrosis.

Pneumoconioses

The term *pneumoconiosis,* originally coined to describe the nonneoplastic lung reaction to inhalation of mineral dusts encountered in the workplace, now also includes diseases induced by organic as well as inorganic particulates and chemical fumes and vapors. A simplified