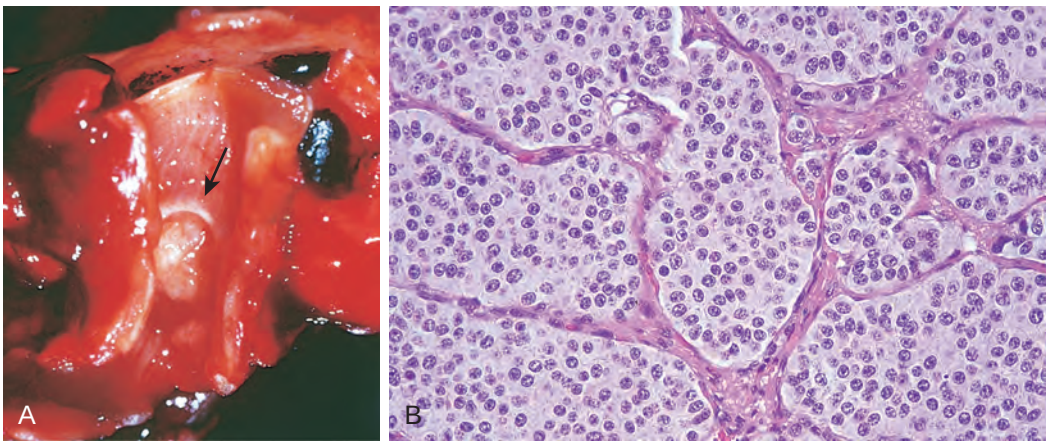

Figure 15-47 Bronchial carcinoid. **A,** Carcinoid growing as a spherical mass *(arrow)* protruding into the lumen of the bronchus. **B,** The tumor cells have small, rounded, uniform nuclei and moderate amounts of cytoplasm. (Courtesy Dr. Thomas Krausz, Department of Pathology, The University of Chicago, Pritzker School of Medicine, Chicago, Ill.)

amount of eosinophilic cytoplasm (Fig. 15-47*B*). Typical carcinoids have fewer than two mitoses per 10 high-power fields and lack necrosis, while atypical carcinoids have between two and 10 mitoses per 10 high-power fields and/or foci of necrosis. Atypical carcinoids also show increased pleomorphism, have more prominent nucleoli, and are more likely to grow in a disorganized fashion and invade lymphatics. On electron microscopy the cells exhibit the dense-core granules characteristic of other neuroendocrine tumors and, by immunohistochemistry, are found to contain serotonin, neuron-specific enolase, bombesin, calcitonin, or other peptides.

Clinical Features. The clinical manifestations of bronchial carcinoids emanate from their intraluminal growth, their capacity to metastasize, and the ability of some of the lesions to elaborate vasoactive amines. Persistent cough, hemoptysis, impairment of drainage of respiratory passages with secondary infections, bronchiectasis, emphysema, and atelectasis are all by-products of the intraluminal growth of these lesions.

Most interesting are functioning lesions capable of producing the classic *carcinoid syndrome*, characterized by intermittent attacks of diarrhea, flushing, and cyanosis. Approximately, 10 % of bronchial carcinoids give rise to this syndrome. Overall, most bronchial carcinoids do not have secretory activity and do not metastasize to distant sites but follow a relatively benign course for long periods and are therefore amenable to resection. The reported 5-year survival rates are 95% for typical carcinoids, 70% for atypical carcinoids, 30% for large cell neuroendocrine carcinoma, and 5% for small cell carcinoma, respectively.

Miscellaneous Tumors

Lesions of the complex category of benign and malignant mesenchymal tumors, such as inflammatory myofibroblastic tumor, fibroma, fibrosarcoma, lymphangioleiomyomatosis, leiomyoma, leiomyosarcoma, lipoma, hemangioma, and chondroma, may occur but are rare. Benign and malignant hematopoietic tumors, similar to those described in other organs, may also affect the lung, either as isolated lesions or, more commonly, as part of a generalized disorder. These include Langerhans cell histiocytosis, non-Hodgkin and Hodgkin lymphomas, lymphomatoid granulomatosis, an unusual EBV-positive B cell lymphoma, and low-grade extranodal marginal zone B-cell lymphoma (Chapter 13).

A lung *hamartoma* is a relatively common lesion that is usually discovered as an incidental, rounded radio-opacity *(coin lesion)* on a routine chest film. Most are, solitary, less than 3 to 4 cm in diameter, and well circumscribed. Pulmonary hamartoma consists of nodules of connective tissue intersected by epithelial clefts. Cartilage is the most common connective tissue, but there may also be cellular fibrous tissue and fat. The epithelial clefts are lined by ciliated columnar epithelium or nonciliated epithelium and probably represent entrapment of respiratory epithelium (Fig. 15-48). The traditional term *hamartoma* is retained for this lesion, but it is in fact a clonal neoplasm associated with chromosomal aberrations involving either 6p21 or 12q14-q15.

Lymphangioleiomyomatosis is a pulmonary disorder that primarily affects young woman of childbearing age. It is

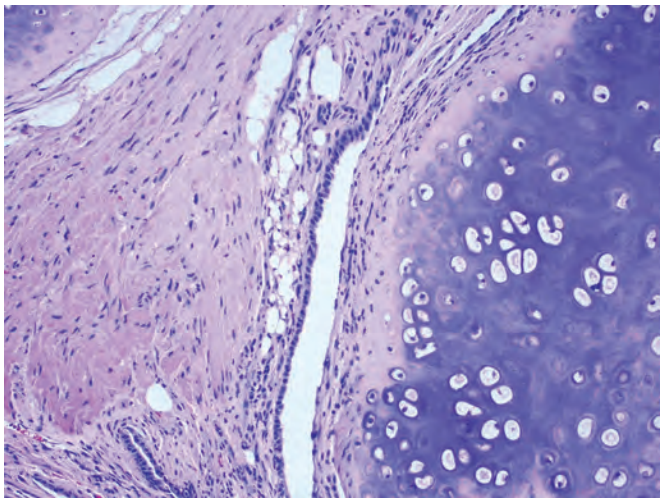

Figure 15-48 Pulmonary hamartoma. There are islands of cartilage, fat, smooth muscle, and entrapped respiratory epithelium.