The patient is a 39-year-old white man who had progressive dyspnea for several years. Recently, at another hospital, he was told he had lung cysts, and right thoracotomy was performed. The present admission resulted because of the sudden onset of acute pleuritic pain in his left chest. A chronic cough, present for three years, yielded foul-smelling, tenacious sputum, occasionally blood-tinged.

Chest roentgenograms obtained seven years earlier were reviewed and appeared normal. PPD 1 intradermal test was negative. Sputum analyses for malignant cells were negative.

*From the Veterans Administration Hospital.
Diagnosis: HISTIOCYTOSIS X

Figure 1 reveals multiple large thin-walled cysts containing varying amounts of fluid and air. At necropsy, numerous huge pulmonary cysts, up to 10 cm. in diameter, were present. Some were filled with viscid greenish-brown material; others with a semi-solid substance. The bronchial tree contained masses of yellowish semi-solid material which filled the lumen like polyps (Fig. 2).

Microscopic examination revealed massive infiltration of the bronchial walls by lipid-filled histiocytes and large collections of cholesterol crystals. The cystic cavities in the lungs were lined by epithelium surrounded by fibrous tissue. They contained cholesterol crystals and lipid histiocytes. The adjacent lung tissue was normal except for edema fluid and early broncho-pneumonia. The axillary nodes showed similar involvement. No other organs were affected.

Discussion

Lichtenstien has proposed the term histiocytosis X to include several related disorders of the reticulo-endothelial system, viz.: Letterer-Siwe’s disease, Hand-schuller-Christian’s disease and eosinophilic granuloma. The disease has also been termed pulmonary xanthomatosis, eosinophilic granuloma of the lung, and “honeycomb” lung. The etiology is obscure. The common microscopic feature is infiltration of tissue by large numbers of histiocytes.

The disease may appear in many organ systems as well as the lungs. However, in no reported instance has histiocytosis X appeared primarily in the lung and later disseminated through the osseous and visceral structures. In the present case, both the pulmonary and lymphatic systems were involved.

The typical roentgen appearance in pulmonary histiocytosis X consists of bilateral disseminated reticulo-nodular densities averaging up to 0.5 cm. in diameter. Extensive cystic changes may be present, but these are generally small and irregular. They have been characterized as “honeycomb-

Figure 2a: Resected lung specimen.

Figure 2b: Cut section of resected lung.

References


