Diagnosis: Congenital Bronchogenic Cyst

The roentgenogram of the chest (Fig 1) demonstrates a large, oblique, homogeneous shadow with a sharp border in the right lower lobe. No calcification is seen within the lesion.

Thoracotomy revealed a 3x5x2 cm thick-walled cystic mass filled with mucoid material deep within the right lower lobe. It showed no abnormal bronchial communication and had no systemic arterial supply. The mediastinal lymph nodes were not enlarged. Lobectomy was performed.

Histologically, the cyst was lined with respiratory epithelial cells, resembling those of bronchial walls. Surrounding the cyst were areas of fibrosis containing cartilage, bone, and nerve tissue.

Bronchogenic cysts may be either solitary or multiple. They are thin-walled cavities of varying size and lie within normal pulmonary tissue. Histologically, the cyst wall consists of an epithelial layer of ciliated columnar cells and elements of the bronchial wall.

Two types of bronchogenic cyst are known, closed and open.1 The closed cyst has no communication with the bronchial tree, is distended, and contains stagnant fluid. As long as no complication intervenes, these cysts are discovered incidentally. Their rate of growth depends on two factors: the surface dimensions of the included mucosa and the strength of the cyst wall. Atelectasis and infection may develop as a result of compression, and are an indication for surgical intervention.2 An open cyst results from communication with a bronchus and secondary infection of the lumen usually occurs, giving rise to the clinical picture of a chronic abscess.

The case under discussion was a closed cyst. Its shape and position suggested intralobar sequestration, a vascular shadow, or benign tumor or cyst.

References

THE EPONYMS AND VAGARIES OF SARCOIDOSIS

Hutchinson in 1875 first described this type of skin lesion and called it Mortier's maladvy after the name of his patient. The second case of this category was reported by Besnier as lupus pernio in 1889. Ten years later, Boeck referred to the disease as multiple benign sarcoid but renamed it benign miliary lupoid in 1905. Heerfordt is credited with the description (in 1909) of uveoparotid fever as a clinical variety of sarcoidosis. Juengling in 1911 observed bone changes due to sarcoidosis and classified them as osteitis tuberculosa multiplex cystica. Schaumann recommended the expressive term benign lymphgranulomatosis for this disease. Of course, no one would call it the Hutchinson-Besnier-Boeck-Heerfordt-Juengling-Schaumann syndrome in lieu of sarcoidosis. The latter is known as one of the most protan and capricious clinical entities. It may involve any of the tissues and organs of the body, with the exception of the suprarenal glands. In reference to diseases of the chest, the following are of special interest: sarcoidosis of the hilar and mediastinal lymph nodes, the lung, the heart and the esophagus. Pulmonary pathologic changes vary from nodular lesions of 1–5 mm in diameter or much larger to honeycomb lung, bronchiectasis, middle lobe syndrome, extensive fibrosis, compensatory emphysema, multiple bullae and rarely, cavity formation. Decreased alveolar diffusing capacity results from specific alveolocapillary block. In some instances, there is discrepancy between the degree of pulmonary dysfunction and respective radiologic findings. Severe lung damage leads to cor pulmonale in about eight per cent of cases. Granulomatous lesions of sarcoidosis of the heart may be localized in the conduction system, right and/or left ventricular wall, interventricular septum, papillary muscles, epicardium and rarely in the pericardium. Even so, no symptoms are present in about one-third of these cases. In others, A-V dissociation is frequent. Complete heart block is observed in one-third of patients with cardiac sarcoidosis. Corresponding ECG changes are noted. In other cases, arrhythmias, frequently paroxysmal, are observed. ECG may appear bizarre, nonspecific and unstable. Sudden death occurs in about 17 per cent of patients with myocardial sarcoidosis. Involvement of the esophagus consists of mucosal and submucosal specific changes associated with fibrosis and ulceration. The etiologic agent of sarcoidosis is unknown. The characteristic feature of the latter is noncaseating granuloma with epithelioid cells. Langhans-type and foreign body giant cells. Asteroid bodies and laminated Schaumann bodies may be seen in the giant cells. Spontaneous recovery takes place in patients with hilar involvement only, in 80–90 per cent, with hilar and pulmonary disease in over 50 per cent, with pulmonary infiltration alone in over 40 per cent. Incidences with subacute onset have a favorable prognosis. The opposite holds true of cases of the chronic type. Mortality rate varies from 5.5 to 28 per cent. It is substantially higher in women than in men. All this being so, there is urgent need for concerted research for the solution of the enigma of sarcoidosis.

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