The Langerhans histiocytes have a close association with parent early origin. The possible role of these cells in histiocytes
previous foamy macrophages nucleus and numerous empty cytoplasmic vesicles, which probably contained lipid. Epithelium (Ep) lines alveolar spaces (A), and fibrous stroma of dense collagen (C) is observed in septal interstitium. Patent capillaries are not seen (× 6,000).

Although it is difficult to reconstruct the events which ultimately result in cyst formation in eosinophilic granuloma, our observations suggest that the initial lesion is an interstitial accumulation of cells, which includes the distinctive Langerhans histiocyte. This infiltrate appears to be associated with, and may be responsible for, the destruction of the parenchyma and the consequent formation of cysts. The cysts with walls demonstrating dense accumulations of inflammatory cells probably are of recent origin. As they mature, the inflammatory infiltrate is replaced by fibroblasts until finally the walls are comprised solely of fibrous tissue. Inasmuch as the parenchymal peripheral to the cysts also undergoes destruction, this tissue may coalesce with the cyst walls. The Langerhans histiocytes have a close association with collagen and reticulin deposits in the cyst walls of apparent early origin. The possible role of these cells in early fibrogenesis remains to be defined.

REFERENCES
1 Heppleston AG: The pathology of honeycomb lung. Thorax 1:77-93, 1956

Granular Cell Myoblastoma of the Bronchus: A New Case, 12-Year Followup Report, and Review of the Literature* 

Ferenc L. Korompai, M.D.;** Robert J. Aue, M.D.;† Arthur C. Beall, M.D., F.C.C.P.;§ and S. Donald Greenberg, M.D., F.C.C.P.||

A case of granular cell myoblastoma of the bronchus is added to the 42 already reported in the English literature. The extensive spread of the tumor along the bronchus is an unusual feature. A 12-year followup on a case previously reported demonstrates cure following adequate resection. From a review of the literature we concluded that this tumor is found most often in the fourth and fifth decade of life, has equal sex incidence and generalized distribution throughout the bronchial tree. The best results follow lung-saving procedures such as bronchotomy, wedge or sleeve resection of the involved bronchus.

Granular cell myoblastoma is a rare benign lesion in the tracheobronchial tree that gives rise to bronchial obstructive symptoms and hemoptysis. The tumor as originally described by Abrikossoff1 was considered to be of muscular origin, but currently neural derivation is favored.2 When granular cell myoblastoma involves the major airways its identification by bronchoscopy and biopsy poses no problem. Reported treatment modalities have ranged from endoscopic resection to pneumonectomy and, in two instances, radiotherapy (Table 1). Most of the extensive pulmonary resections were performed because of bronchiectasis and lung destruction secondary to the obstructing tumor.

Table 1—Treatment Modalities Used in the Reviewed Series

<table>
<thead>
<tr>
<th>Procedure</th>
<th>No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Biopsy only</td>
<td>3</td>
</tr>
<tr>
<td>Endoscopic resection</td>
<td>9</td>
</tr>
<tr>
<td>Bronchotomy</td>
<td>1</td>
</tr>
<tr>
<td>Sleeve or wedge of bronchus</td>
<td>5</td>
</tr>
<tr>
<td>Sleeve with pulmonary resection</td>
<td>3</td>
</tr>
<tr>
<td>Segmentectomy</td>
<td>2</td>
</tr>
<tr>
<td>Lobectomy</td>
<td>13</td>
</tr>
<tr>
<td>Pneumonectomy</td>
<td>5</td>
</tr>
<tr>
<td>Radiotherapy</td>
<td>2</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>43</td>
</tr>
</tbody>
</table>

*From the Cora and Webb Mading Department of Surgery, and Departments of Medicine and Pathology, Baylor College of Medicine, Houston, Texas.
**Instructor in Surgery.
†Assistant Professor of Medicine.
‡Professor of Surgery.
§Associate Professor of Pathology.
||Supported in part by the USPHS (HE-05435 and HE-05387) and the Lester Kamin Respiratory Pathology Research Fund.

Reprint requests: Dr. Beall, Baylor College of Medicine, 1200 Mournard Avenue, Houston 77025

CHEST, 66: 5, NOVEMBER, 1974
Case Report

A 36-year-old Negro woman (cigarette smoker) was admitted to the hospital with a three-week history of right anterior chest pain which was initially throbbing, but later became pleuritic. Gross hemoptysis occurred for 48 hours before admission. Increasing cough and yellow sputum production was present for six months. Decreased breath sounds and scattered rales were noted anteriorly on the right. Chest x-ray film findings showed right middle lobe consolidation. Sputum findings were negative for acid-fast bacilli, and routine cultures grew normal respiratory flora.

The diagnosis of granular cell myoblastoma was established by bronchoscopy and biopsy (Fig 1, 2). The tumor was light gray to pale pink, firm, nodular and was occluding the middle lobe orifice and bulging into, but apparently not involving, the mucosa of the bronchus intermedius.

At thoracotomy the right middle lobe was atelectatic, and the lobar bronchus and adjacent bronchus intermedius were thickened. Right middle lobectomy was performed, with sleeve resection of the bronchus intermedius. Although grossly normal, on frozen section both proximal and distal resection margins contained tumor. Because of proximity of positive margins to the remaining lobar bronchial orifices, resection of the lower lobe and right main bronchus was performed. The right upper lobe bronchus was reimplanted into the trachea. The remaining upper lobe expanded well, and the patient was discharged on the tenth day after operation.

Discussion

One of the first studies to refute the theory that granular cell tumors arose as myoblastomas from embryonal muscle rests was that of Pearse3 who found the cytoplasmic granules of the “myoblastoma” to contain lipid, protein and glycol groups and proposed that the lesion represented granular degeneration in perineural fibroblasts. In 1962, in a histochemical and electron-microscopy study, Fisher and Wechsler2 provided information that also indicated the derivation of these lesions from Schwann cells. Sobel and Churg,4 in 1964, reported additional histochemical and ultrastructural studies of granular cell tumors. They concluded that the idea of neural derivation of granular cell myoblastoma was an attractive one and most likely correct. Electronmicroscopic study of the granular cell tumor in this case revealed similar osmiophilic cytoplasmic granules to those reported by Fisher and Wechsler2 and Sobel and Churg; however, myelin figures were infrequent (Fig 2). Tissue frozen section and routine light microscopic study of the bronchial tumor in this case showed the granular cells to be closely packed and of varying shapes (Fig 1). The nuclei were oval to round. No mitotic figures were seen. The cytoplasm of the tumor cells contained eosinophilic staining poorly defined granules. Similar histologic features were found in both the biopsy and the lobectomy specimens.

This case provided an opportunity to obtain a 12-year followup on a patient with bronchial granular cell myoblastoma reported previously from this institution.8 She was a 32-year-old Negro woman whose tumor involved the right lower lobe bronchus and was treated with right middle and lower lobectomy on May 12, 1961. Subsequently, she had two granular cell myoblastomas of the skin removed, one from the left chest wall in February, 1962 and one from the right arm in March, 1962. Her current physician in California was contacted, and he reported no recurrence of either skin or bronchial lesions.

A review of the reported cases of bronchial granular cell myoblastomas5-13 showed that the median age was 40 years, with the greatest incidence in the fifth decade. The sex distribution was even. The location of the tumors revealed generalized distribution throughout the bronchial tree.

Local excision such as endoscopic resection, bronchotomy, or bronchial wedge resection with bronchoplasty has proven possible in most patients. This case report illustrates a lesion that appeared localized at bronchoscopy, but due to endobronchial spread of the tumor found at operation, an extensive resection was required. Frozen section studies of the resection margins were valuable in carrying out the proper surgical treatment.

Figure 1. Light micrograph showing closely packed granular cells of bronchial tumor. Nuclei are regular, with evenly dispersed chromatin. Cytoplasm is abundant and finely granular. Cell borders are indistinct. There are no mitoses. Appearance of tumor was similar in frozen tissue preparations seen at operation (H&E stain, x 875).

Figure 2. Electronmicrograph of granular cell tumor from bronchus. Note prominent and varying sized closely packed cytoplasmic cytosomes, some of which are osmiophilic. There is more or less even distribution between clear vacuoles and osmiophilic lipid granules. Tumor cells have finely dispersed chromatin and single prominent nucleolus (uranyl acetate & lead citrate stain, x 12,000).
Mitral Valvotomy in a Patient with Dextrocardia and Situs Inversus

Allan M. Lansing, M.D.,** and E. L. W. Scofield, M.D.†

Mitral valvotomy was performed in a 45-year-old woman with mirror-image dextrocardia, situs inversus and acquired mitral stenosis. The literature was reviewed and a total of 15 reported cases was found, with surgical treatment in 4. Closed mitral valvotomy has been the treatment in all cases to date, and was successful in each. An open-heart approach was perfectly feasible in our case, but was reserved for the future when valve replacement might be required.

**Professor of Surgery, University of Louisville School of Medicine, Health Sciences Center, Louisville, Ky.
†Resident in Thoracic and Cardiovascular Surgery, University of Louisville School of Medicine.
This study was supported in part by USPHS grant no. 5T12HE05504 and The Louisville Medical Research Foundation.

Dextrocardia with situs inversus is an uncommon defect, and the association of this congenital lesion with acquired valvular heart disease is even rarer. A recent experience with this combination led to our review of the literature and case report.

**Case Report**

The patient was a 45-year-old woman with a history of acute rheumatic fever 22 years before, at which time she was found to have a heart murmur and dextrocardia. Over the subsequent nine-month period the patient was admitted to the hospital three more times for treatment of the rheumatic fever. She was then asymptomatic until one year prior to her admission for cardiac evaluation on Oct. 11, 1971. During this period she had noted a gradually progressive increase in dyspnea on exertion, easy fatigability, difficulty in doing her housework, paroxysmal nocturnal dyspnea, bilateral pedal edema, and two-pillow orthopnea. At the time of admission the patient was considered to have cardiac functional disease class 3, but had been taking no cardiac medication.

Physical examination revealed a moderately obese woman with a blood pressure of 130/90 mm Hg and normal sinus rhythm at a rate of 88/min. The positive physical findings were limited to the heart. The apex beat was in the fifth right intercostal space at the midclavicular line. There were no palpable heart sounds or thrills, and there was no evidence of ventricular hypertrophy. The first heart sound was moderately accentuated as was the pulmonary component of the second heart sound. There was no third or fourth heart sound. An opening snap was heard over the entire precordium, which the phonocardiogram demonstrated .06 sec after the aortic closure, and there was a grade 2/6 diastolic rumbling murmur at the apex, with presystolic accentuation. There was no systolic murmur. The chest x-ray film revealed dextrocardia and situs inversus, a cardiothoracic ratio of 0.64/29.6, left atrial enlargement, and prominence of the central and upper lobe vessels (Fig 1). The electrocardiogram was consistent with dextrocardia and also showed...

**Figure 1. Note dextrocardia, enlarged left atrium, and prominence of pulmonary vascularity.**