present case appears to be unique in the sense that two-dimensional echocardiography permitted direct dynamic visualization of small emboli moving freely in the right-sided chambers, toward the pulmonary artery. Clinical and postmortem evidence was thereafter obtained to corroborate the diagnosis raised by echocardiography. We ruled out a hypothesis of microcavitations created by intravenous therapy because the infusion was very slow to produce images with the intensity observed in this case. We have recently infused peripherally normal subjects as well as patients with overt heart failure, at rates of up to 3 ml/min, without seeing any images like those documented in this case. Only when the infusion was done through catheters placed in the superior vena cava have we been able to detect microcavitations during slower infusion rates. In the same way, the echodense images visualized in this case appear to be more intense than the microbubbles observed in tricuspid regurgitation.

Although it is difficult to ascertain the diagnostic value of two-dimensional echocardiography, in the clinical context of suspected thromboembolism, we believe that the specificity and sensitivity of such noninvasive methods should be measured by further studies. It is possible that the finding of mobile emboli in the right cardiac chambers can be useful for immediate assessment of the need of emergency surgery for embolectomy or interruption of the inferior vena cava, in selected critically ill patients, thus avoiding the risks of invasive approaches such as angiography.

Pulmonary Artery Obstruction due to Malignant Fibrous Histiocytoma*

Brian W. Carlin, M.D.,† and Kenneth M. Moser, M.D., F.C.C.P.

Obstruction of the pulmonary artery can be due to a variety of conditions, including chronic thromboembolic disease, neoplasia, and fibrosing mediastinitis. Differentiation can be difficult. We report a case of malignant fibrous histiocytoma causing intravascular obstruction and include a review of the literature.

The differential diagnosis of chronic thromboembolic obstruction of major pulmonary arteries includes rare instances of neoplasms originating in these arteries.1 Most such tumors are diagnosed post mortem.2 We report herein the findings in a patient who presented with total obstruction of the left main pulmonary artery, in whom the diagnosis was made ante mortem and in whom resection was performed.

Case Report

A 68-year-old man was well until September 1985, when dyspnea on exertion developed. In October a paroxysmal nonproductive cough and daily evening temperature elevations appeared. Symptoms briefly resolved with antibiotic therapy and then recurred. A chest x-ray film in December 1985 demonstrated fullness in the aortopulmonary window and left hemidiaphragmatic elevation. Pulmonary function tests revealed the following: forced expiratory volume in one second (FEV1), 1.51 L (43 percent of predicted); forced vital capacity, 3.01 L (60 percent of predicted); and carbon monoxide diffusing capacity (Dsb), 14.6 ml/min/mm Hg (46 percent of predicted). Bronchoscopic findings were unremarkable; brushings and washings revealed only atypical cells. A computerized tomogram of the chest suggested a filling defect in the left main pulmonary artery. A gallium scan showed abnormal uptake in the left peribilar region. A perfusion scan (Fig 1) showed no perfusion to the left lung and several subsegmental defects in the right lung. A digital-subtraction pulmonary angiogram showed complete occlusion of the left main pulmonary artery, with a normal right main pulmonary artery. An echocardiogram was normal.

When the patient was referred to our institution in early January, his symptoms included shortness of breath, a nonproductive cough, and an 11.3-kg (25-lb) loss of weight since September. He was afebrile on admission and remained so. The findings from physical examination were unremarkable. The patient was mildly anemic (hematocrit reading, 32 percent). A chest x-ray film and perfusion scans were unchanged. Arterial blood gas analysis disclosed the following values: pH, 7.45; arterial oxygen pressure, 62 mm Hg; and arterial carbon dioxide tension, 41 mm Hg. An electrocardiogram showed left anterior fascicular block. Pulmonary function tests showed the following: FEV1, 1.66 L/sec (50 percent of predicted); ratio of residual volume over total lung capacity, 57 percent; and Dsb, 11.9 ml/min/mm Hg (49 percent of predicted). Oxygen saturation did not decline with exercise at 2 mph for six minutes. The results of impedance plethysmography of the legs were normal. Cardiac catheterization disclosed pulmonary arterial pressure of 34/20 mm Hg (mean 26 mm Hg), a pulmonary arterial wedge pressure of 10 mm

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movement. Chest and abdominal computerized tomographic scans disclosed no evidence of metastatic disease. A left mediastinotomy was negative. We elected to explore the patient's pulmonary arteries through a median sternotomy. On opening the left main pulmonary artery, we encountered thrombotic material obstructing it; however, as dissection was extended, whitish quite friable material was encountered which extended into the left upper lobe artery and its segmental branches. The left descending pulmonary artery was occluded with what appeared to be organized thrombus. The right main pulmonary artery and its lobar and segmental branches were normal. A saphenous venous bypass of the right coronary artery was performed.

Pathologic examination of the material removed disclosed proximal organized thrombus distal to which was a highly pleomorphic spindled neoplasm with frequent atypical mitoses and scattered multinucleate cells. The diagnosis of intravascular malignant fibrous histiocytoma was made. There was no evidence of invasion into the left pulmonary arterial wall, a small portion of which (just beyond the origin of the occlusion) was biopsied. The material removed from the left descending pulmonary artery was organized thrombus.

Some weeks later, after a further detailed evaluation for metastatic disease was negative, the patient underwent a left pneumonectomy. Pathologic examination of the lung removed revealed an intravascular tumor in the left upper lobe segmental branches, with a single area of invasion through a segmental pulmonary artery into the pulmonary parenchyma. Mediastinal lymph nodes were, as at the first operation, normal. Ten months after surgery, the patient was doing well, without evidence of recurrence.

**Discussion**

This patient presented with many of the historical, physical, and laboratory features associated with chronic thrombotic obstruction of major pulmonary arteries; however, he was found to have a unique cause of pulmonary artery obstruction, an intravascular malignant fibrous histiocytoma. Organized thrombus had developed proximal (left main pulmonary artery) and distal (left descending pulmonary artery) to the tumor.

Shortness of breath, progressive dyspnea on exertion, a nonproductive cough, and the lack of a clear-cut history of venous thromboembolism are presentations common to both thromboembolic and neoplastic obstruction of the pulmonary artery; however, our patient's history of loss of weight and fever was atypical for patients with chronic thromboembolism.

Roentgenographically, decreased pulmonary vascular markings or volume loss can be seen in both diseases, but the presence of a hilar mass, infiltrate, or parenchymal nodules suggests tumor involvement. Filling defects on the perfusion scan are universally present in both states. Many patients with neoplastic obstruction of the pulmonary artery are treated for venous thromboembolism, and the diagnosis of a pulmonary artery tumor is rarely considered ante mortem.

At surgery the two diseases can resemble each other, and only histologic examination can provide the diagnosis. In many cases, as in ours, thrombus formation occurs in concert with the tumor itself.

Malignant fibrous histiocytoma was initially characterized by O'Brien and Stout as a group of tumors with a common origin from tissue histiocytes. The diagnosis has achieved greater frequency in recent years, and many cases previously diagnosed as pleomorphic rhabdomyosarcoma or undifferentiated fibrosarcoma are now classified as malignant fibrous

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**FIGURE 1.** Perfusion scan (anterior view) showing no perfusion to left lung and several subsegmental defects in right lung.

Hg, and a cardiac index of 3.0 L/min/sq m. Bilateral pulmonary angiography (Fig 2) revealed total obstruction of the left pulmonary artery shortly beyond its origin; there was no evidence of thromboembolic disease of the right pulmonary artery or its branches. Coronary angiography revealed a 90 percent stenosis of the right coronary artery.

Fluoroscopic examination demonstrated normal diaphragmatic movement.

**FIGURE 2.** Angiogram of left pulmonary artery showing total obstruction distal to bifurcation.
Table 1—Clinicopathologic Observations in 20 Patients with Malignant Fibrous Histiocytoma of the Lung

<table>
<thead>
<tr>
<th>Reference</th>
<th>Year</th>
<th>Age (yr)</th>
<th>Surgery</th>
<th>Radiation Therapy</th>
<th>Chemotherapy</th>
<th>Metastases</th>
<th>Duration of Life</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kern et al(1)</td>
<td>1977</td>
<td>1, M, 53</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>Cerebral</td>
<td>5 mo ...</td>
</tr>
<tr>
<td>Bedrossian et al(3)</td>
<td>1979</td>
<td>3, M, 51</td>
<td>+ (\dagger)</td>
<td>-</td>
<td>-</td>
<td>Liver</td>
<td>3 mo ...</td>
</tr>
<tr>
<td>Chowdhury et al(4)</td>
<td>1980</td>
<td>4, F, 52</td>
<td>+ (\ddagger)</td>
<td>-</td>
<td>-</td>
<td>Cerebral; renal</td>
<td>5 mo ...</td>
</tr>
<tr>
<td>Misra et al(5)</td>
<td>1981</td>
<td>5, M, 45</td>
<td>+ (\dagger)</td>
<td>-</td>
<td>-</td>
<td>Cerebral</td>
<td>5 mo ...</td>
</tr>
<tr>
<td>Sajjad et al(6)</td>
<td>1981</td>
<td>6, F, 65</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>Cerebral</td>
<td>12 mo ...</td>
</tr>
<tr>
<td>Mills et al(7)</td>
<td>1982</td>
<td>7, F, 60</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>Sternal</td>
<td>8 mo ...</td>
</tr>
<tr>
<td>Lee et al(8)</td>
<td>1984</td>
<td>8, M, 62</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>Liver; pericardium</td>
<td>6 mo ...</td>
</tr>
<tr>
<td>Ros et al(9)</td>
<td>1984</td>
<td>12F, 14F</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>Atrial</td>
<td>11 mo ...</td>
</tr>
<tr>
<td>Case report(10)</td>
<td>1985</td>
<td>15, F, 46</td>
<td>+ (\dagger)</td>
<td>-</td>
<td>-</td>
<td>Intraoral</td>
<td>2 mo ...</td>
</tr>
<tr>
<td>Venn et al(11)</td>
<td>1986</td>
<td>16, F, 32</td>
<td>+</td>
<td>-</td>
<td>+</td>
<td>-</td>
<td>15 mo ...</td>
</tr>
<tr>
<td></td>
<td></td>
<td>17, M, 62</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>60 mo ...</td>
</tr>
<tr>
<td></td>
<td></td>
<td>18, M, 33</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>Atrium</td>
<td>18 mo ...</td>
</tr>
<tr>
<td></td>
<td></td>
<td>19, M, 62</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>120 mo ...</td>
</tr>
<tr>
<td></td>
<td></td>
<td>20, F, 61</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>60 mo ...</td>
</tr>
</tbody>
</table>

*Involvement of pulmonary artery found. \(\dagger\) Patient had bilateral nodules, one of which was resected. \(\ddagger\) Resection not performed, only biopsy of pulmonary mass. \(\dagger\) Tumor found at embolectomy; subsequent surgery revealed no tumor. \(\dagger\) Patient died of congestive heart failure without evidence of tumor recurrence. \(\dagger\) Neither characteristics of patients nor therapy discussed in article.

This tumor is believed to be the most common soft tissue sarcoma of adult life. While primarily found in the extremities, abdominal cavity, and retroperitoneal region, 10,12 20 cases of malignant fibrous histiocytoma involving the pulmonary parenchyma primarily have been described (Table 1\(1^{18-8}\)). In 15 cases, surgical resection was performed. Survival ranged from three months to ten years, with two long-term survivors (five years and ten years) in the group with resection. In no prior case did the tumor appear to arise within the pulmonary artery.

This case illustrates several points. First, pulmonary artery tumors can mimic many of the signs, symptoms, and findings associated with chronic thrombotic obstruction of the pulmonary artery. Secondly, a high index of suspicion can lead to early surgery and, hopefully, an improved outcome.

ACKNOWLEDGMENT: We thank Dr. Ben Bridges of the University of Texas Tyler Medical Center for his kindness in providing follow-up data about this patient.

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