symptoms for another 15 months prior to cardiac catheterization and subsequent surgery.

This case illustrates the importance of considering postoperative ostial stenosis in the differential diagnosis of angina following an operation which involves cannulation of the coronary arteries. This diagnosis should be considered regardless of the procedure or the length of time between the operation and the onset of symptoms. The angiogram in these patients usually shows left main coronary artery stenosis, although occasionally right coronary, circumflex or left anterior descending artery involvement has been noted. Due to the effect of left main stenosis on survival, an aggressive diagnostic and therapeutic approach should be employed in these patients to reduce subsequent morbidity and mortality.

REFERENCES


Pulmonary Lymphangiomyomatosis Associated with Tuberous Sclerosis*

Treatment with Tamoxifen and Tetracycline-pleurodesis

Carlos M. Luna, M.D.; Ricardo Gené, M.D.; Enrique C. Jolly, M.D.; Nelly Nahmod, M.D.; Héctor A. DeFranchi, M.D.; Guillermo Patiño, M.D.; and Boris Elsner, M.D.

A 44-year-old woman was seen with the clinical and histologic picture of lymphangiomyomatosis syndrome. She also had dermatologic and neurologic stigmata of tuberous sclerosis. After the development of a recurring chylothorax, she had a downhill course unresponsive to dietary, bronchodilator, corticosteroid and progestosterone therapy. In an open lung specimen, the search for steroid receptor for estrogen was positive. The discovery in this case of an estrogen receptor represents important evidence for establishing an association between tuberous sclerosis and lymphangiomyomatosis. Tamoxifen therapy and tetracycline pleurodesis were successful in stopping the progressive course and controlling the chylothorax.

Pulmonary lymphangiomyomatosis (PLM) is a rare disease that predominantly affects women in their reproductive years. It results from a benign proliferation of the smooth muscle in lungs and other organs. Cystic changes and honeycombing may be seen in the lungs of patients with more advanced disease. The result is an interstitial and obstructive lung disease.

The chief complaint of these patients is usually dyspnea. Pneumothorax, hemoptysis and chylothorax are common complications.

Diffuse lung disease clinically and histologically resembling PLM has also been described in tuberous sclerosis. There is some doubt whether PLM is one of the hamartomatous lesions known to be associated with tuberous sclerosis.

There is no successful therapy for PLM. Most patients die from respiratory insufficiency within ten years of diagnosis.

The primary occurrence of this disease in women of reproductive age had suggested a hormonal influence in the etiology of PLM.

Attempts to treat this condition included: corticosteroids, androgens, progestosterone, castration and discontinuation of exogenous estrogens. The possible usefulness of tamoxifen has been suggested and trials of that drug have been recommended.

We report a patient in whom expressions of both disorders coexisted. We treated with tamoxifen, with the addition of the use of tetracycline-pleurodesis to control a recurring chylothorax.

CASE REPORT

A 44-year-old woman was first seen in August, 1981 complaining of breathlessness of two years’ duration.

She denied consuming oral contraceptives. Remarkable physical findings included fibrous papules on the face, shaligreen patches on her back and few scattered rales at lung bases. There was no family history of tuberous sclerosis. The serum creatinine was 1.4 mg/dl. She had average high intelligence (Weschler Bellevue test) and phakomatous retinal lesions certified by an angiofluoresceinography. A computerized axial tomogram revealed cerebral calcifications. Ultrasonography demonstrated bilateral renal angiomyolipomas and gamma camera scanning showed multiple negative shadows and deformations in both kidneys. There was no difference in the estimated contribution of each kidney to the total renal function. Pulmonary function tests disclosed severe obstructive ventilatory defect and hypoxemia (Table I). A transbronchial lung biopsy performed through a fiberoptic bronchoscope showed smooth muscle arranged in interlacing bundles that were characteristics of PLM.

The pharmacologic treatment included: oral aminophylline 220 mg and inhaled salbutamol 200 µg, gid; from October, 1981 to October, 1982, prednisone treatment in progressively tapering doses, from 60 mg per day to 8 mg every other day; and medroxyprogesterone acetate 1 gr per month intramuscularly from December, 1981 to July, 1982.

In April, 1982 she developed a left chyloous pleural effusion (Fig 1)
and her clinical condition became worse. Repeated thoracocentesis and dietary treatment did not resolve the chylothorax. In October, 1982 a limited left thoracotomy and open lung biopsy, and subsequently tetracycline pleurodesis, were performed.

Sections of the lung biopsy showed irregular smooth muscle proliferation located mainly in the wall of pulmonary vessels. There was disruption of the elastic lamella and reduction of the lumen (Fig 2).

Focal smooth muscle hyperplasia was seen in the wall of some bronchi and alveolar walls. Intrapulmonary lymphatics were dilated, but not involved by smooth muscle proliferation. A mild degree of emphysema was present, but fibrosis was absent.

Estrogen receptor was determined by a charcoal-dextran method on a sample of the lung. The value was 32 f mol/mg-protein (positive = greater than 3 f mol/mg protein). Tamoxifen therapy (20 mg per day) was then started. The patient's respiratory status and pulmonary function improved (Table 1).

In April, 1983 she developed a serious spontaneous hemorrhage inside the right angiomyolipomatous kidney; after a nephrectomy, she recovered uneventfully. In August, 1984 serum creatinine level was 1.2 mg/dl, there was an objective improvement of the fibrous papules of the face, she had evidence of arrested disease, and her exercise tolerance improved.

**DISCUSSION**

Patients such as this (female of childbearing age with pulmonary lesion, chylothorax, and clinical stigmata of tuberous sclerosis), provide evidence that PLM is indeed a **forme fruste** of tuberous sclerosis. 3,5

The two rare entities produce identical spectra of pulmonary clinical, roentgenologic and histologic changes and occur almost exclusively in women. 3,5,9-10

Two groups of investigators 6,12 emphasized the absence of pulmonary lymphatic involvement in patients with tuberous sclerosis. However, in one study the ultrastructural morphology was found to be absolutely identical in tuberous sclerosis and PLM. 7

Castration as a means of therapy has been associated with a stable respiratory picture in PLM. 6,14 As a logical conclusion of this observation, the use of antiestrogens has been suggested. 6,7

The occurrence of objective regression or stabilization with ablation and hormonal therapy in patients with PLM has insinuated that steroid receptor determination might be clinically relevant. 15,16 Progesterone and estrogen receptors

---

**Table 1—Serial Pulmonary Function Values before and after Pleurodesis and Tamoxifen Therapy**

<table>
<thead>
<tr>
<th>Date of Studies</th>
<th>Age of Patient</th>
<th>Basal FVC (L)</th>
<th>Basal FEV (L)</th>
<th>Basal PaO₂ (mm Hg)</th>
<th>Basal PaCO₂ (mm Hg)</th>
<th>Post Exercise Basal FVC (L)</th>
<th>Post Exercise Basal FEV (L)</th>
<th>Post Exercise Basal PaO₂ (mm Hg)</th>
<th>Post Exercise Basal PaCO₂ (mm Hg)</th>
<th>BUN (mg/dl)</th>
<th>Creatinine in Blood (mg/dl)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oct 1981</td>
<td>44</td>
<td>1.600</td>
<td>1.760</td>
<td>0.880</td>
<td>1.016</td>
<td>1.050</td>
<td>1.030</td>
<td>32</td>
<td>38</td>
<td>0.980</td>
<td>1.650</td>
</tr>
<tr>
<td>Jan 1982</td>
<td>45</td>
<td>2.350</td>
<td>2.600</td>
<td>0.930</td>
<td>1.030</td>
<td>1.070</td>
<td>1.050</td>
<td>32</td>
<td>38</td>
<td>0.980</td>
<td>1.650</td>
</tr>
<tr>
<td>Jul 1982</td>
<td>45</td>
<td>1.650</td>
<td>1.760</td>
<td>0.800</td>
<td>1.016</td>
<td>1.050</td>
<td>1.030</td>
<td>32</td>
<td>38</td>
<td>0.980</td>
<td>1.650</td>
</tr>
<tr>
<td>Sep 1982</td>
<td>45</td>
<td>1.930</td>
<td>2.150</td>
<td>0.980</td>
<td>1.090</td>
<td>1.070</td>
<td>1.050</td>
<td>32</td>
<td>38</td>
<td>0.980</td>
<td>1.650</td>
</tr>
<tr>
<td>Dec 1982‡</td>
<td>45</td>
<td>2.340</td>
<td>2.780</td>
<td>0.980</td>
<td>1.090</td>
<td>1.070</td>
<td>1.050</td>
<td>32</td>
<td>38</td>
<td>0.980</td>
<td>1.650</td>
</tr>
<tr>
<td>Sep 1983‡</td>
<td>46</td>
<td>2.700</td>
<td>2.610</td>
<td>0.920</td>
<td>1.090</td>
<td>1.070</td>
<td>1.050</td>
<td>32</td>
<td>38</td>
<td>0.980</td>
<td>1.650</td>
</tr>
<tr>
<td>Aug 1984‡</td>
<td>47</td>
<td>2.350</td>
<td>2.600</td>
<td>0.830</td>
<td>0.920</td>
<td>1.070</td>
<td>1.050</td>
<td>32</td>
<td>38</td>
<td>0.980</td>
<td>1.650</td>
</tr>
</tbody>
</table>

*Percentage of normal value.
†Exercise on bicycle ergometer: 90 sec without load, 60 sec with 300 kg/min.
‡Post pleurodesis, during tamoxifen therapy.
were found in PLM tissue;\textsuperscript{15,16} this has never been demonstrated, to our knowledge, in tuberous sclerosis.

There are two previously published reports related to the use of tamoxifen in PLM.\textsuperscript{16,17} Both women had severe respiratory insufficiency and the drug was ineffective in arresting the course of the disease, but tamoxifen was given at a very late stage. One of them had cor pulmonale and the other, who had high level of estrogen receptor, was seriously ill and on mechanical ventilation at the start of therapy.

The administration of pharmacologic doses of tamoxifen during 24 months and the use of tetracycline pleurodesis was associated with improvement of some clinical and laboratory manifestations and stabilization of the respiratory picture in our patient. Both measures seem to be effective therapies for this rare pulmonary disease and the response to tamoxifen is probably related to the presence of an estrogen receptor.

Finding an estrogen receptor in this patient is new evidence that supports the association between PLM and tuberous sclerosis.

ACKNOWLEDGEMENT. We are indebted to Dr. J. C. Hogg for reviewing the histologic material.

REFERENCES

5 Kitzsteiner KA, Mallen RG. Pulmonary lymphangiomyomatosis; treatment with castration. Cancer 1980; 46:2248-49

Massive Hemoptysis Associated with Foreign Body Removal*

J. Richard Rees, M.D., F.C.C.P.\textsuperscript{+}

Exsanguinating hemoptysis accompanied removal of an endobronchial foreign body in a 12-year-old child. Preparations to treat this complication should be made prior to removal of any foreign body of prolonged sojourn in the tracheobronchial tree.

The most common serious complication accompanying bronchoscopy for removal of foreign bodies in the tracheobronchial tree is cardiac arrest due to asphyxia.\textsuperscript{1-8} From a review of the literature, massive hemorrhage accompanying bronchoscopy for foreign body removal is an unusual complication.

CASE REPORT

A 12-year-old white girl presented to the hospital with fever, chest pain and hemoptysis. An admitting chest x-ray film (Fig 1) demonstrated an infiltrate in the right lower lobe and a foreign body in the right main stem bronchus. The patient gave a vague history of having swallowed a bullet seven years previously. The foreign body, an intact .22 short caliber cartridge casing, was removed easily through a rigid endoscope. Immediately after removal of the foreign body, massive hemorrhage into the right main stem bronchus ensued, which was then tamponaded with cotton and gauze. The patient was placed in the left lateral decubitus position and an emergency right thoracotomy was performed. The patient then sustained cardiopulmonary arrest. The right mainstem bronchus was opened proximal to the tamponade and a considerable amount of old blood was aspirated from the left main stem bronchus as successful cardiac massage was carried out.

Postoperatively, the patient developed adult respiratory distress syndrome plus a bronchopleural fistula requiring prolonged mechanical ventilation and the construction of an Elosser flap. She subsequently recovered, underwent plastic repair of the chest wall with two artificial ribs and has done well.

DISCUSSION

In their monumental work on foreign bodies in air and food passages, Jackson and Jackson described 343 patients with prolonged sojourn of foreign bodies in the air passages.\textsuperscript{1} These same authors were able to remove most of these foreign bodies transbronchoscopically and, in most instances, noted resolution of the underlying inflammatory process in the

*From the University of Utah College of Medicine, Salt Lake City.\textsuperscript{+}Assistant Clinical Professor of Surgery. Reprint requests: Dr. Rees, 425 East 5350 South, Ogden, Utah 84405