of contents for decompression is insufficient.

The gastric mucosal lacerations described have their analogies additionally in forceful emesis and therapeutic epigastric compression. In 1929 Mallory and Weiss brought to our attention the former phenomenon with their description of 15 patients believed to have had gastroesophageal lacerations with hemorrhage secondary to vomiting. Heimblich introduced a maneuver for the choking victim in 1974, and less than 18 months later the medical literature bore a report of gastric rupture complicating that life-saving maneuver.

No previous reports of fatal hemorrhage from gastric lacerations related to CPR can be identified. Particularly with the raging popularity of bystander initiation of CPR, it is important to recognize that the mechanics of resuscitation itself are potentially morbid, even fatal. Because of the evidence that gastric lacerations may be common, and now the present report of fatal hemorrhage, attention to the details of CPR and its follow-up are imperative. It would serve the resuscitator well to attempt proper mouth-to-mouth ventilation, perhaps with early tracheal intubation, provide for gastric decompression with nasogastric suction, and insure appropriately placed and timed chest compression. However, despite our awareness and intentions, it can be anticipated that there will be subsequent reports of gastroesophageal laceration with hemorrhage and perforation with CPR. Evidence of upper GI bleeding peririsu citation should be approached aggressively in light of the current information.

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The Association of Progressive Multifocal Leukoencephalopathy and Sarcoidosis*

Mark A. Rosenbloom, M.D.; and Dean F. Uphoff, M.D.

A 59-year-old woman had a right homonymous hemianopsia, memory impairment for five months, a nonenhancing area in the left parieto-occipital region on CT scan, and bilateral reticulonodular infiltrates on chest x-ray film. Lung biopsy findings were consistent with sarcoidosis, a clinical diagnosis of CNS sarcoidosis made, and prednisone therapy begun. She deteriorated neurologically and died. At autopsy characteristic histologic and electron microscopic features of progressive multifocal leukoencephalopathy (PML) were found. We conclude that this and other cases demonstrate an association of PML and sarcoidosis and that steroid treatment is not a precondition. We also suggest an aggressive diagnostic approach in evaluating sarcoidosis with atypical neurologic deficits.

Patients with sarcoidosis demonstrate immunologic abnormalities such as anergy, a depression of systemic cell-mediated immunity, and a hyperactivity of the B cell system. Despite abnormalities in cell-mediated immunity the only well-documented infectious complication of sarcoidosis is aspergillosis in areas of the lung with cystic changes. However, in a recent review on the neurologic manifestations of sarcoidosis, Delaney noted six cases of progressive multifocal leukoencephalopathy (PML), a viral infection, that have been reported in patients with sarcoidosis. Our purpose is to present a patient demonstrating both well documented PML and sarcoidosis and to review the previously reported cases.

CASE REPORT

A previously healthy 59-year-old woman was admitted for evaluation of persistent visual and memory difficulties during the preceding five months. Approximately four months previously, the patient was involved in an automobile accident. She suffered no apparent injury, but medical evaluation revealed a right homonymous hemianopsia. Two months after the accident, a CT scan showed a very small hypodense area in the left parieto-occipital lobes, without projection to the cortical surface. A brain scan was normal.

There was no history of rashes, fevers, headaches, seizures, focal weakness, shortness of breath, smoking, or weight loss. She was taking no medication and had had no prior hospitalizations.

Physical examination at admission showed her to be well-nourished, in no respiratory distress, and afebrile. Her pulse rate was 70 beats/min, blood pressure 120/70 mm Hg, and respiration 14/min. Examination of the eyes revealed normal pupils and papillary reflexes, with intact extraocular movements. The neck was normal, and the breasts had no masses. The lungs were clear, and the heart was normal. No abdominal masses or hepatosplenomegaly were noted. There were no rashes or lymphadenopathy. Neurologic examination showed her to be alert and well-oriented, but unable to subtract serial 75 and having difficulty in number recall. She had a right homonymous hemianopsia. Motor, sensory, and cerebellar testing results were normal. Deep tendon reflexes were symmetric.

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and 2+, and no pathologic reflexes were elicited. Chest x-ray film showed extensive bilateral reticulonodular infiltrates.

Contrast CT scan showed a nonenhancing irregular area involving primarily the white matter in the left parieto-occipital region. The lesion had increased in size compared with the previous CT scan.

Laboratory findings included the following: hematocrit readings, 38.9 percent; leukocyte count, 4,100/mm3, with 57 percent neutrophils, 7 percent bands, 2 percent eosinophils, 20 percent lymphocytes, and 14 percent monocytes; and the ESR was 59 mm/hr. Serum electrolytes, BUN, and creatinine and glucose levels were normal. The alkaline phosphatase, serum transaminase, lactic dehydrogenase, and bilirubin levels also were normal. Serum protein electrophoresis was normal, and angiotension-converting enzyme was 56 µl/ml (normal 0 to 30). A lumbar puncture revealed an opening pressure of 120 mm, 120 RBCs/mm3 and 0 WBCs/mm3; the protein was 13 mg/dl; glucose, 72 mg/dl, and all cultures, India ink preparation, and AFB stain were negative. Skin tests with intermediate-strength PPD, Candida, and mumps were negative.

Clinical Course

An open lung biopsy was performed, revealing histologic abnormalities consistent with sarcoidosis (Fig 1). Although the neurologic presentation was atypical, the presumptive diagnosis was CNS sarcoidosis, and prednisone therapy, 60 mg daily, was started and continued for ten weeks. The patient was readmitted because of progressive confusion. A repeated lumbar puncture result was normal, and a CT scan showed a new nonenhancing area on the opposite side. A brain biopsy of the initial left occipital lobe lesion was performed and showed pallor of myelin in the gyral white matter and evidence of myelin destruction in the form of perivascular macrophages. No intranuclear inclusions in oligodendroglia or atypical astrocytes were found; electron microscopy examination failed to show viral particles.

The presumptive diagnosis was PML. She lapsed into a coma and died shortly thereafter.

Autopsy

Autopsy findings included multiple small nodular foci of fibrosis in the lungs which on microscopic study appeared to be the residua of the sarcoid granulomas seen on the lung biopsy specimen months earlier. The spleen also showed numerous fibrotic nodules consistent with healed sarcoid granulomas. Other viscera and lymph nodes were unremarkable.

Coronal sections of the brain showed extensive demyelination of the white matter at the biopsy site, which on microscopy failed to show evidence of active inflammation or features of PML. There were numerous small foci of demyelination becoming confluent in the right temporal lobe white matter, and smaller, fewer demyelinated areas of the frontal lobe and the cerebellar white matter. On

![Figure 1. Biopsy of lung showing sarcoid granulomas (H & E objective x 6).](image1)

![Figure 2. Demyelinated area of autopsy brain showing inflammatory reaction with macrophages and large intranuclear inclusions (arrows). (H & E objective x 16.)](image2)

![Figure 3. Electron micrograph of formalin-fixed autopsy brain showing spherical intranuclear viral particles 32 nm in diameter characteristic of Papova virus. (Original magnification x 4000; inset magnification x8000.)](image3)

Discussion

At the time of her initial presentation, our patient had pulmonary and neurologic abnormalities that required explanation. She was taking no medication and had had neurologic symptoms for approximately five months prior to hospitalization. Once the diagnosis of advanced pulmonary sarcoidosis was made by examination of open lung biopsy specimen and chest x-ray film abnormalities, the etiology of the CNS pathology was paramount. CNS sarcoidosis is known to occur in about 5 percent of patients with sarcoidosis, although autopsy studies suggest the incidence is probably higher. The neurologic manifestations in this patient had several features atypical of CNS sarcoidosis. As mentioned in several reviews, CNS sarcoidosis has a predilection for the base of the brain and tends to occur early in the disease, usually in patients younger than 40 years old. The most common neurologic manifestations are therefore cranial neuropathies, basilar meningitis, and hypothalamic or pituitary involve-
Sarcoidosis presenting as an intracerebral mass is uncommon. However, among eight patients reviewed by Norwood and Kelly who presented with CNS sarcoid mimicking a mass lesion, four of them had homonymous hemianopsia. Associated CSF abnormalities are frequently found, and Delaney noted an increased protein or pleocytosis (predominantly lymphocytes) in approximately 70 percent of patients with CNS sarcoidosis. The atypical neurologic features in our patient included her age, the advanced stage of her pulmonary sarcoidosis, the normal lumbar puncture, and the anatomic location of her lesion. In spite of the atypical neurologic findings, it was thought that CNS sarcoidosis was likely and potentially treatable, and prednisone therapy was begun. After the patient deteriorated neurologically during the ten weeks of steroid therapy, PML seemed even more likely. Although the brain biopsy specimen revealed nonspecific demyelination, the autopsy data confirmed PML as the neurologic disease.

This patient illustrated many typical features of PML. Her clinical course of progressive neurologic deficits with deteriorating mental status, culminating in coma and death, is characteristic of PML. The CT findings were similar to those described by Carroll et al—a lucent, nonenhancing area with scalloped lateral borders, involving primarily the white matter. The CFS findings in PML are usually normal or show a mild increase in protein. Although the etiology of this disease is known to be the Papova virus (JC virus or SV 40), the treatment is unsatisfactory. Aside from sporadic reports, the disease is still almost uniformly fatal.

Although PML is most commonly seen in patients with chronic lymphocytic leukemia, Hodgkin's disease, and non-Hodgkin's lymphoma, some association with sarcoidosis has been suggested in the previous literature. In a review of 83 cases of PML, Richardson noted that five of these patients also had sarcoidosis. A total of seven patients have been reported to have PML and sarcoidosis although the diagnosis of sarcoidosis is questionable in two of them. In 1955, Christensen and Fog described the first patient in which these two diseases occurred concurrently. The patient presented with a right homonymous hemianopsia and with reading and writing difficulties. A chest x-ray film showed diffuse infiltrates, and the patient began receiving ACTH therapy for a clinical diagnosis of sarcoid. He deteriorated neurologically and died about four months after his presentation. At autopsy, noncaseating granulomas with Schaumann's and asteroid bodies were found in the lung. Microscopic examination of the brain revealed several characteristic features of PML, although this was three years before Avstrom described PML as a distinct entity. This patient, similar to the one we reported, concurrently had advanced

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Source</th>
<th>Age, Sex at Onset of CNS Disease</th>
<th>Path. Diagnosis of Sarcoid</th>
<th>CNS Pathology</th>
<th>Other Diseases</th>
<th>Steroids at Time of CNS Presentation</th>
<th>Time From Onset of CNS Disease to Death</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Christensen, Fog (1955)</td>
<td>59 M</td>
<td>Autopsy—noncaseating granulomas with Schaumann's and asteroid bodies in lung</td>
<td>Autopsy—demyelination with monstrous multinucleated astrocytes</td>
<td>None</td>
<td>No</td>
<td>5 mo</td>
</tr>
<tr>
<td>2</td>
<td>Richardson* (1961)</td>
<td>47 F</td>
<td>—</td>
<td>Autopsy—reviewed by Richardson, typical lesions and cell changes</td>
<td>None</td>
<td>?</td>
<td>4 mo</td>
</tr>
<tr>
<td>3</td>
<td>Loken, Refsum, Jacobsen (1962)</td>
<td>47 M</td>
<td>Autopsy—noncaseating granulomas in lung, lymph nodes, and spleen</td>
<td>Autopsy—demyelination, giant astrocytes, enlarged hyperchromatic oligodendroglia nuclei</td>
<td>None</td>
<td>No</td>
<td>8 mo</td>
</tr>
<tr>
<td>4</td>
<td>Davies, Hughes, Oppenheimer (1973)</td>
<td>46 M</td>
<td>Autopsy—mostly noncaseating granulomas in mediastinal and abdominal lymph nodes, liver, and spleen</td>
<td>Autopsy—EM-viral particles in oligodendroglial cells</td>
<td>None</td>
<td>No</td>
<td>4 mo</td>
</tr>
<tr>
<td>5</td>
<td>Marriott et al (1975)</td>
<td>52 F</td>
<td>By history of 14 years diagnostic procedure not described</td>
<td>Brain biopsy—demyelination in white matter, oligodendroglial inclusions, viral particles in oligodendroglial on EM, viral culture</td>
<td>None</td>
<td>7.5 mg prednisone for 7 years</td>
<td>Patient alive 2½ years after cytarabine therapy, with significant resolution of neurologic symptoms</td>
</tr>
</tbody>
</table>

*A personal communication from J. Hallervorden to E. Richardson.
pulmonary sarcoidosis and evidence of brain disease prior to steroid therapy.

Marriott in 1976 described a case in which both diagnosis of PML and sarcoidosis seemed unequivocal. A 52-year-old woman receiving daily prednisone for seven years for pulmonary sarcoidosis presented with dysphagia, right hemiplegia, and impaired concentration. A brain biopsy was performed, confirming the diagnosis of PML and allowing identification of the JC virus. After five days of cytarabine therapy (2mg/kg/day), a slight improvement was noted. The patient continued to improve neurologically and ultimately was able to return to work as a nursing tutor. She represents one of the very few cases of PML in which a favorable therapeutic response occurred. Walker suggested that because patients with sarcoidosis have some residual immunity, addition of cytarabine (an inhibitory viral agent) might allow the host resistance to regain control and halt progression of disease.

The remaining cases of PML and sarcoidosis are summarized in Table 1.

In conclusion, the association of sarcoidosis and PML in our patient is unequivocal. This case and the cases reviewed support the contention that although rare, an association of sarcoidosis and PML does exist. It has been stated that opportunistic infections occurring in patients with sarcoidosis are a consequence of steroid therapy. The association of PML and sarcoidosis in this patient and in the literature suggests that use of steroids is not a precondition. We suggest that PML should be considered in any patient thought to have CNS sarcoidosis, particularly if the CSF is normal. In addition, we suggest an aggressive diagnostic approach in evaluating cases of sarcoidosis with atypical neurologic deficits.

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Isolated Aortic Origin of Right Pulmonary Artery

Report of a Case with Special Reference to Pulmonary Vascular Disease in the Left and Right Lungs

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Morphometric study of the pulmonary vasculature was performed on lung biopsy specimens from a one-year-old girl who underwent anatomic repair of isolated aortic origin of the right pulmonary artery. Medial hypertrophy of small pulmonary arteries in the right lung was much less remarkable than that in the left lung. In contrast, intimal lesions in the right lung were much more advanced than those in the left lung. Fully oxygenated blood in the right pulmonary artery might suppress medial hypertrophy in response to high pressure and thin media fail to protect intima from high pressure, resulting in severe intimal lesions. This situation in the right lung resembles that in complete transposition of the great arteries.

Isolated aortic origin of the right pulmonary artery is a rare congenital heart anomaly, and surgical intervention has not led to favorable results. Principal among the reasons for such poor surgical results is the early development of pulmonary vascular disease. However, few detailed studies of the pulmonary vascular lesions in this disease with its characteristic hemodynamics have been reported.

Recently we experienced anatomic repair of a one-year-old

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