The diagnosis of this rare disease is often delayed even in the presence of pulmonary symptoms. The most common presenting symptom is pneumothorax, followed by dyspnea with exercise. The average delay in diagnosis is 44 months after the onset of symptoms. Our diagnosis occurred only 2 months after the presenting abdominal pain and prior to any pulmonary involvement.

Radiography is usually the first step in diagnosis of LAM and includes chest radiographs, chest CT, and more recently HRCT. Lymphangioleiomyomatosis is associated with chest radiograph findings of pleural effusion, pneumothorax, and interstitial lung disease without loss of lung volumes. In more advanced stages, lung volumes tend to increase. High-resolution CT allows visualization of small cystic spaces not always seen with chest radiography or conventional CT and can be an earlier diagnostic tool than pulmonary function tests. In our patient, neither chest radiography nor chest CT visualized the diffuse parenchymal cysts that HRCT clearly did. Diagnostic distinctions have been described among LAM, emphysema, and histiocytosis. As shown in Figure 3, small diffuse thin-walled cysts randomly located in all lung fields are typical of LAM. This finding in a young, nonsmoking woman is nearly pathognomonic for LAM. Transbronchial biopsy specimens or open-lung biopsy specimens are required for a definitive diagnosis.

Treatment recommendations vary for this little understood disease. Antiestrogen therapy with tamoxifen and oophorectomy have shown varied success. Progesterone therapy using medroxyprogesterone acetate was first described by McCarty et al and is perhaps one of the more effective treatments. In one of the largest studies to date on LAM (82 patients), Taylor et al concluded no difference in the rate of progression of the disease with regard to the type of therapy with 25 of 32 patients surviving after 8 years. In their study, however, the only patients with improvements of symptoms were in the medroxyprogesterone group. We hope for a better prognosis in our patient since therapy was instituted relatively early.

REFERENCES

Hypersensitivity Pneumonitis due to Native Birds in a Bird Ringer*

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A bird fancier who had only been exposed to native birds, mostly owls, developed a severe hypersensitivity pneumonitis with a very insidious onset.

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*BFL=bird fancier's lung

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BIRD FANCIER'S LUNG (BFL) IS USUALLY CAUSED BY EXPOSURE TO THE ANTIGENS OF PIGEONS AND BUDGERIGARS.1 OTHER BIRDS REPORTED AS A CAUSE OF THE DISEASE INCLUDE CANARIES, PARROTS, DOVES, CHICKENS, TURKEYS, DUCKS, AND GESE. IN THE LITERATURE WE FOUND TWO CASES IN WHICH BFL WAS CAUSED BY CONTACT WITH NATIVE BIRDS.2 WE REPORT A CASE OF BFL, WITH A VERY INSIDIOUS ONSET, DUE TO EXPOSURE TO NATIVE BIRDS, MAINLY OWLS.

CASE REPORT


The patient had been ringing native birds since 1957, 100 to 500 yearly. The work was done during the spring. After 1976, he had been ringing annually also 100 to 200 owls. In 1992, the number of ringed owls was 200. During the same period, he also had kept in his home in wintertime many native birds, which usually had been found injured. Small birds were kept in his living rooms and bigger birds, eg, owls, in the basement. Since 1989, he had kept only owls and had during three winters housed eagle owls (Bubo bubo) for 1 to 4 months on each occasion. In May 1992, he had housed a Tengmalm’s owl (Aegolius funereus) in the basement for 2 weeks. He had had no exposure to pigeons, chickens, turkeys, ducks, or geese or caged birds such as budgerigars or canaries.

On hospital admission, his chest radiograph revealed severe linear and reticular, mainly fibrotic changes (Fig 1). Retrospectively, already in 1982, minor changes in the chest radiograph were visible and these changes had gradually increased with time. The erythrocyte sedimentation rate was slightly elevated (31 mm/h, normal <20 mm/h). Hemoglobin, blood leukocyte count, and serum angiotensin-converting enzyme were normal. Spirometry showed slight restriction (FEV1, 3.15 L, 72 percent of predicted, and FVC, 3.71 L, 66 percent of predicted). No bronchodilator response in spirometry was found. The pulmonary diffusing capacity was severely impaired (Dco, 7.5 ml/min/mm Hg, 24 percent of predicted). Arterial blood gas analysis revealed moderate hypoxemia (PaO2, 58 mm Hg; PaCO2, 38 mm Hg). Precipitating antibodies as assessed by the gel precipitation technique were found to pigeon serum and droppings, to budgerigar serum and droppings, and to hen serum. Owl serum was not available for testing.

Bronchoscopy revealed a normal macroscopic appearance. Bronchial biopsy specimen was normal and the transbronchial biopsy specimen was not representative. Thoracic high-resolution computed tomography showed nonspecific fibrosis with honeycombing; no hilar adenopathy was found. Because of severity of the disease, corticosteroid treatment with 48 mg of methylprednisolone was started in August 1992. In September 1992, a thoracoscopic lung biopsy3 was performed. At the time of the lung biopsy, the daily dose of methylprednisolone was 32 mg. The lung biopsy specimen showed evident fibrosis with a mild interstitial infiltrate of lymphocytes and plasma cells. Along bronchiolar and alveolar walls, there were clusters of histiocytes, some showing foamy cytoplasm. Numerous scattered giant cells were also found (Fig 2). The changes were compatible with chronic hypersensitivity pneumonitis.

The corticosteroid treatment was continued for 6 months with decreasing doses. There was a slight improvement in the chest radiograph but major fibrotic changes remained. Pulmonary diffusing capacity rose to 41 percent of predicted at the 7-month follow-up and there was also a slight improvement in spirometry. The patient showed signs of clinical improvement but remained dyspneic during exercise. He agreed to terminate his exposure to birds.

FIGURE 2. Lung biopsy specimen showing fibrosis and interstitial chronic inflammatory cells with a giant cell in the middle consistent with chronic hypersensitivity pneumonitis (van Giesen stain, original magnification ×66).

PHOTOGRAPH 1. Chest radiograph at hospital admission showing linear and reticular changes consistent with chronic hypersensitivity pneumonitis.

DISCUSSION

We found in the literature only one report with two cases of BFL due to exposure to native birds.5 In that report, of 459 cases of BFL from former East Germany, two were caused by native birds. The types of exposure were not accurately described, but the birds involved were sikikins, finches, and goldfinches. In that report, other cases of BFL were caused by pigeons, budgerigars, hens, and canaries. Our patient fulfilled the criteria for hypersensitivity pneumonitis.5 He had been exposed to native birds during his bird-ringing activities and when taking care of native birds in his own home. During the most recent years, he...
had been exposed mainly to owls and his symptoms markedly increased during the last three springs at the time of bird ringing. Exposure to molds or other causative antigens of hypersensitivity pneumonitis had not occurred. Thus, the source of antigens most likely was owls; nevertheless, antigens for serologic testing were not available.

The patient had serum antibodies to pigeon, budgerigar, and hen antigens, even though he had had no contact with these birds. It is evident that these antibodies were caused by cross reactivity between avian antigens. Cross reactivity between sera of patients with BFL and antigens of native birds without known exposure of the patients to these antigens has been reported.6

The clinical course of the disease was insidious and the patient had no fever. Diagnosis of chronic BFL may be very difficult and the disease may be mistaken for an idiopathic pulmonary fibrosis. An accurate diagnosis in our case was important to convince the patient of the need to strictly avoid any future exposure to birds.

In conclusion, native birds may cause BFL, which may have an insidious onset. This potential hazard should be considered in bird fanciers who are bird ringers or who take care of injured native birds.

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A 32-year-old homosexual man presented with a 1-month history of progressive dyspnea, fever, cough, and weight loss (15 kg). His medical history was unremarkable. On examination, he was thin (45 kg) with a temperature of 38°C and oral candidiasis. The respiratory rate was 25 beats/min. Chest examination revealed rales at lung bases. Room air blood gas revealed the following: PaO₂ 71 mm Hg; PaCO₂ 29 mm Hg; and pH 7.50. Chest radiograph demonstrated diffuse reticular interstitial infiltrates and bilateral, diffuse thin-walled air cysts (Fig 1). Human immunodeficiency virus (HIV) serologic study was positive at the time of hospital admission. The CD4 count was 42/mm³. Bronchoalveolar lavage fluid examination yielded P. carinii, and no other organisms. Treatment with intravenous trimethoprim (0.96 g/d) and sulfamethoxazole (4.8 g/d) started on the same day. Six days later, the patient developed right-sided pneumothorax. Lung re-expansion was obtained following the placement of three chest tubes with evidence of a persistent air leak. A CT scan confirmed the presence of generalized cystic air pulmonary lesions (Fig 2). At day 15, the patient developed a contralateral

We describe a case of air cyst lesions in an AIDS patient suffering from Pneumocystis carinii pneumonia. This case is unique because these lesions were generalized to both lungs and initially well tolerated. Pathologic examination revealed extensive tissue invasion by P. carinii. The prognosis was complicated by bilateral pneumothoraces. Surgical right pleurodesis allowed lung re-expansion but did not prevent recurrence of fatal contralateral pneumothorax. (Chest 1994; 106:1271-72)

Bilateral diffuse interstitial infiltrates have been recognized as the most common radiographic manifestation of Pneumocystis carinii pneumonia (PCP) in patients with AIDS.1 Other radiographic patterns include single or multiple air cyst pulmonary lesions.1,2 We report the first case (to our knowledge) of bilateral generalized air cyst lesions on computed tomographic (CT) scan in an AIDS patient without previous opportunistic infection.

CASE REPORT

Generalized Air Cysts Complicated by Fatal Bilateral Pneumothoraces in a Patient With AIDS-Related Pneumocystis carinii Pneumonia*

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FIGURE 1. Initial posteroanterior chest radiograph demonstrates bilateral interstitial disease and multiple air cysts in a patient with PCP.