Waldenstrom’s Macroglobulinemia Presenting as a Pulmonary Mass*

David Major, M.D.;** Mae H. Meltzer, M.D.;† Alexander Nedwich, M.D.;§ Brian Hayes, M.D.,§ and Wilbur W. Oaks, M.D., F.C.C.P.||

We describe an unusual case of Waldenstrom’s macroglobulinemia. It demonstrates the rare presentation of this disease with a pulmonary mass and the therapeutic response to plasmaphoresis and long-term chlorambucil therapy during a greater than two-year followup.

The typical presentation of Waldenstrom’s macroglobulinemia includes the symptoms of fatigue, weight loss, epistaxis, and decreased visual acuity associated with anemia, hepatosplenomegaly, lymphadenopathy and retinal hemorrhage.1,2 A rare case of Waldenstrom’s macroglobulinemia presenting primarily as a pulmonary mass was recently seen at Hahnemann Medical College and Hospital.

CASE REPORT

A 52-year-old white man first became ill in September 1968. He was hospitalized at that time at another institution with a right-sided pyopneumothorax treated by thoracotomy with pleural decortication and drainage of the empyema. A lung biopsy was reported as pseudolymphoma. His recovery was uneventful. In October, 1970 he was hospitalized at Hahnemann Medical College and Hospital because of increasing dyspnea of two months’ duration.

Physical Examination

Physical examination revealed the following pertinent findings: dyspnea at rest, dullness to percussion, scattered moist rales, rhonchi, tubular breathing and a pleural rub over the right posterior upper lung field, liver palpable 4 cm and spleen 6 cm below the costal margins.

Laboratory Studies

Significant findings were Hct 38 percent, WBC count 10,900 with 7 percent atypical lymphocytes and 5 percent lymphocytoid plasma cells. Chest x-ray film revealed generalized consolidation of the right upper lobe and portions of the right lower and right middle lobe with compression of the right mainstem bronchus and narrowing of the right upper lobe bronchus (Fig 1). Protein electrophoresis—monoclonal gammopathy 5.0 gm percent (Fig 2). Immunoelectrophoresis—IgM. Urine was negative for protein.

Pathology

Bone marrow aspiration revealed hypercellular marrow with a predominance of lymphocytoid plasma cells. The other cellular elements were decreased in numbers. Percutaneous lung biopsy of the right upper lobe was compared to tissue from his previous thoracic surgery. Both revealed the alveolar tissue to be completely obliterated by a marked proliferation of lymphocytoid plasma cells of lymphoreticular origin with prominent nuclei surrounded by a clear cytoplasmic halo. This halo corresponded to sites of PAS diastase resistant positive reaction suggesting areas of accumulation of glycoproteins (Figs 3 and 4).

Hyperviscosity Syndrome

On the 11th hospital day the patient developed decreased

Fig. 2. Serum protein electrophoresis revealing marked monoclonal gammopathy.

Downloaded From: http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/20945/ on 04/08/2017
visual acuity, malaise, weakness and dilated "sausage-like" segmentation of the retinal veins. Serum viscosity was 21 seconds (control 6 sec). Plasmaphoresis (4,000 ml in four days) alleviated the syndrome; chlorambucil was instituted: (8 mg/day x 2 weeks then 4 mg/day).

Follow-up
At three months, the patient was asymptomatic, the liver was not palpable, spleen 3 cm; chest x-ray film: decreased size of mass (Fig 5). At one year, the patient remained asymptomatic with mild leukopenia, spleen not palpable, chest x-ray appearance stable. Chlorambucil was reduced to 2 mg/day. At two years, the patient continues to be asymptomatic. The area of percussion, dullness and tubular breathing has continued to diminish in size. The roentgenographic opacity is now restricted to the right apex and infraclavicular region. Hematocrit is 42 percent. White blood cell count is 5,230 with four atypical lymphocytes and five lymphocytoid plasma cells. The monoclonal gammopathy remains, but is only 2.5 gm percent. The patient is maintained on 2 mg of chlorambucil daily.

DISCUSSION

Waldenstrom's macroglobulinemia first described in 1944 is a lymphoproliferative disorder in which the basic pathologic finding is the proliferation of neoplastic secretory lymphocytoid cells. These cells have been described variously as plasmacytoid lymphocytes, lymphocytoid plasmocytes, plasmalymphocytes or lymphoreticular cells. By electron microscopy these cells reveal endoplasmic reticulum and Golgi structures which characteristically are seen in plasma cells. At autopsy, these cells are most commonly distributed in a diffuse

Figure 3. Needle biopsy of right lung showing obliteration of normal architecture (hematoxylin and eosin, ×30).

Figure 4. Needle biopsy of the right upper lobe of lung, showing marked proliferation of lymphocytoid plasma cells, scattered immature plasma cells, lymphoblasts and a few large cells with prominent nuclei surrounded by a clear halo (hematoxylin and eosin, ×1000).

Figure 5. Posterioanterior chest roentgenogram on Feb. 17, 1971 after three months of chemotherapy with chlorambucil revealing decrease in the size of the pulmonary mass.
scattered pattern involving mainly the bone marrow, liver, spleen and lymph nodes. However, the compact localization of these cells forming tumor masses is a rare occurrence.

It has been well documented that patients with Waldenström's macroglobulinemia5,6 are predisposed to a variety of pulmonary infections. However, tumor formation by the neoplastic cells in the lungs is indeed a rare presentation. McCallister and colleagues7 review of 227 cases did not include any such instance. Furgeson et al8 reported an autopsy case with pulmonary involvement consisting of fine nodular infiltrates, as well as large areas of confluent infiltration of the lungs due to proliferation of plasmacytoid neoplastic cells, similar plasmacytoid cells being seen in the bone marrow. Moeschlin9 also documented a case of pulmonary involvement by antemortem lung biopsy also with marrow involvement. Recently, Rabiner et al10 reported a patient with early bronchial and pulmonary involvement with negative bone marrow. In the few previously reported cases of pulmonary tumor in Waldenström's macroglobulinemia and in the present one, histopathologic findings disclosed the classic spectrum of the neoplastic lymphoproliferative cells, the predominant plasmacytoid plasmaocyte, other types of lymphocytes, plasma cells and primitive reticulum cells. The relation between clinical symptoms and the degrees of serum hyperviscosity in Waldenström's macroglobulinemia have been discussed by Fahey and others.11 The beneficial effect of plasmaphoresis on hyperviscosity syndrome has been known for some time and was well demonstrated in our patient.12 The efficacy of chlorambucil therapy is documented by McCallister and others.7,8,10,13,14 Relapse frequently occurred following discontinuation of chlorambucil with inability in some cases to induce a second remission when the drug is readministered.8 In the present case, somewhat over two years of continuous chlorambucil therapy has resulted in abolition of symptoms, control of the hyperviscosity syndrome, resolution of the hepatosplenomegaly, decrease in the size of the pulmonary mass and disappearance of the funduscopic abnormalities.

References


Pentazocine Addiction causing Bacterial Endocarditis and Pulmonary Embolism*

Najab Ali, M.D.,**, and Tazewell Banks, M.D.†

An emotionally unstable medical technician, without prior history of drug dependence who developed pentazocine addiction, is described. Chronic, unsupervised, intravenous use of crushed tablets of pentazocine caused bacterial endocarditis, pulmonary embolism, and pulmonary edema-like picture. Our patient illustrates that even in the absence of past history of drug dependence, the use of pentazocine should, at best, be avoided in emotionally unstable patients, and patients with easy access to the drug.

Pentazocine (Talwin), a weak narcotic derived from the benzomorphans nucleus, has been described as a potent nonaddictive analgesic.1 Over the past five years, a number of reports have described patients who developed addiction to the drug.2,4 This communication will

*From the Department of Cardiology, Howard University Medical Division, D.C. General Hospital, Washington, D.C.
**Chief of Cardiology, Assistant Professor of Medicine, Howard University Medical Division, D.C. General Hospital.
†Associate Professor of Medicine, Chief Cardiologist, Howard University Medical Division, D.C. General Hospital.