Sjogren Syndrome Presenting as Pulmonary Pseudolymphoma: Report of a Case

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A case of pulmonary pseudolymphoma and Sjogren syndrome is presented. Unusually, the lung involvement preceded the salivary disease by more than two years. The initial gammopathy and abnormal serologies are more consistent with uncomplicated Sjogren syndrome, but were present when the pseudolymphoma was solely apparent.

Sjogren syndrome precedes pseudolymphomatous development in the cases reported in the literature.[1-3] A case report is presented in which pulmonary pseudolymphoma precedes the clinical symptoms of the sicca complex by more than two years. The presence of multiple pulmonary nodules of pseudolymphoma makes this case a rare presentation of Sjogren syndrome.

Case Report

A 40-year-old woman was being seen for a bacterial pneumonia in 1975 when a chest radiograph showed bilateral pulmonary nodules (Figure 1). There was no past pertinent medical history. A hemogram and routine chemistries were normal. Total protein was 10.0 gm/dl with a serum globulin of 6.6 gm/dl. Immunoglobulins (Ig) G and A were increased to 27.0 mg/ml and 10.5 mg/ml, respectively. A normal IgM of 2.80 mg/ml was obtained. Antinuclear antibody and latex fixation tests were not performed (Table 1).

The patient was referred to the UCLA Center for the Health Sciences in July 1976, for further evaluation. Following a normal physical examination, the patient's chest radiograph (Figure 2) revealed an increase in size of the pulmonary nodules but no other abnormalities. Total protein, serum globulin, and immunoglobulin levels were unchanged from 1975. The antinuclear antibody was positive at 1:20 with a speckled pattern. Rheumatoid factor was present at a dilution of 1:10,240 (Table 1). Spirometry was normal. The diffusing capacity was 61 percent of the predicted value. A percutaneous lung biopsy was performed under fluoroscopy and the section histologically revealed a dense interstitial lymphoplasmacytic infiltrate which compressed, but did not invade, bronchioles. Because this biopsy was questioned, a repeat open-lung biopsy was performed. The section confirmed the percutaneous biopsy and demonstrated compression of the bronchioles without invasion (Figure 3). The histologic and radiographic diagnosis of pulmonary pseudolymphoma was made and the patient was followed without treatment.

In August 1977, bilateral parotid and submaxillary gland enlargement and progressive xerostomia and keratoconjunctivitis sicca developed. A Schirmer's test was grossly positive and a labial biopsy was consistent with Sjogren syndrome. Dyspnea was present on exertion. The total protein was 11.2 gm/dl with a serum globulin of 6.4 gm/dl. The IgG level was 73.4 mg/ml with an IgA level of 15.1 mg/ml and IgM level of 2.4 mg/ml. The speckled pattern of antinuclear antibody increased to a titer of 1:160 (Table 1). The rheumatoid factor and chest radiography were unchanged. Diffusing capacity decreased to 51 percent of the predicted value. Steroid therapy was instituted and dyspnea improved; the diffusing capacity increased to 67 percent of predicted value. The chest radiograph remained unchanged while the xerostomia and keratoconjunctivitis sicca resolved. The total protein level decreased to 9.0 gm/dl with a serum globulin level of 4.3 gm/dl. The antinuclear antibody findings became negative and the rheumatoid factor decreased to 1:5120. A labial biopsy following institution of steroid therapy showed mild edema and scattered lymphocytic infiltrates.

Discussion

Pulmonary involvement by pseudolymphoma following Sjogren syndrome has been described.[1-3] In all of the reported cases, the pulmonary pseudolymphomatous infiltrates have appeared following the diagnosis of Sjogren syndrome. This case had pulmonary pseudolymphoma more than two years preceding the onset of Sjogren syndrome. Diffuse interstitial pulmonary fibrosis and pulmonary lymphomas have preceded the onset of Sjogren syndrome in other reports.[3,4]

Extrasalivary lymphoproliferative involvement in Sjogren syndrome by pseudolymphoma, regardless of its site, may be associated with immunoglobulin and serological abnormalities. IgM

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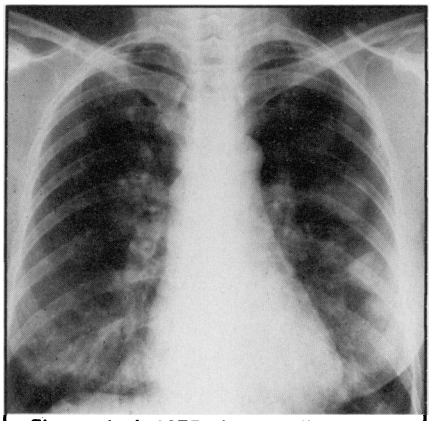

Figure 1. A 1975 chest radiograph showing bilateral, ill-defined, pulmonary nodules.

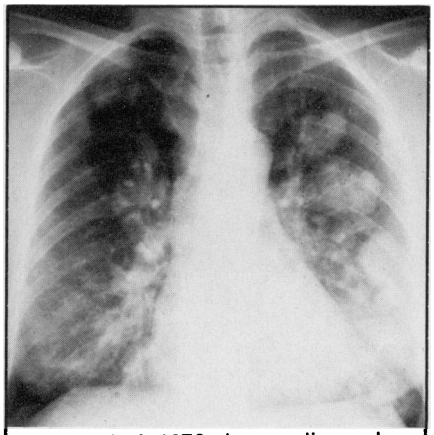

Figure 2. A 1976 chest radiograph revealing increase in size and number of pulmonary nodules.

Table 1. Serum Proteins, Immunoglobulins, and Serologies and Their Changes with Evaluation and Treatment Over a Two-Year Period

	1975	1976	1977	1977 (with treatment)
Total Protein (gm%)	10.0	NC	11.2	9.0
Serum Globulins (gm%)	6.6	NC	6.4	4.3
IgG (mg/ml)	27.0	NC	73.4	—
IgA (mg/ml)	10.5	NC	15.1	—
IgM (mg/ml)	2.8	NC	2.4	—
Antinuclear Antibody	—	1:20	1:160	0
Latex Fixation	—	1:10,240	1:10,240	1:5,120

*NC=no change
—=not performed

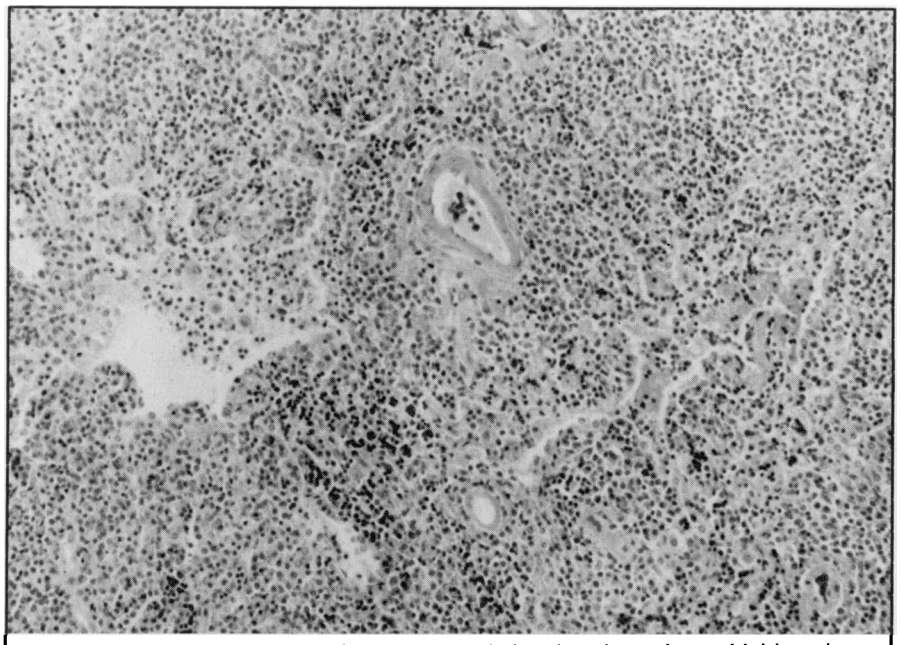

Figure 3. Histologic section of case reported showing dense interstitial lymphoplasmacytic infiltrate compressing but not invading bronchioles (400x).

is characteristically increased while IgG is less often elevated.[2,3] IgA is rarely reported as being raised. Rheumatoid factor is routinely of very high titer.[5] In contrast, characteristic laboratory findings in uncomplicated Sjogren syndrome usually include an antinuclear antibody, high titer rheumatoid factor, and hypergammaglobulinemia.[6] The latter usually consists of an elevated IgG level but IgM and A levels can be increased.[2]

Conclusion

In this patient, the elevated IgG and A and near normal IgM are more consistent with uncomplicated Sjogren syndrome. However, these immunoglobulin levels were elevated for more than two years prior to the diagnosis of Sjogren syndrome. The presence of the increased IgG level and very high titer rheumatoid factor is consistent with pseudolymphoma in Sjogren syndrome while an increased IgA level has rarely been reported. The absence of a high concentration of IgM differs markedly from those cases with pulmonary pseudolymphoma, but is found in uncomplicated Sjogren syndrome. Thus the present case had immunoglobulin and serological abnormalities more consistent with Sjogren syndrome than with pseudolymphoma although the latter temporally preceded the former clinically. The prognosis is indeterminate at the time of this writing.

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