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Pulmonary function abnormalities in Sjögren's syndrome and the sicca complex

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ABSTRACT The frequency of pulmonary involvement in a group of 20 patients with Sjögren's syndrome or the sicca complex was evaluated with pulmonary function studies. In 12 patients pulmonary functional abnormalities were demonstrated. The most common abnormality was airway obstruction. Nine out of 13 patients with the limited variant of the disease (sicca complex) and three out of seven patients with the complete syndrome had abnormal pulmonary function.

Sjögren's syndrome is a chronic autoimmune disorder, characterised by keratoconjunctivitis sicca (dry eyes), xerostomia (dry mouth), and a connective tissue disorder, usually rheumatoid arthritis. The term "sicca complex" is reserved for cases in whom only the first two features are present.

Until recently only isolated cases of pulmonary involvement in patients with Sjögren's syndrome have been reported. Strimlan et al found pulmonary involvement in 9% of patients with Sjögren's syndrome. They showed mainly a restrictive ventilatory defect, low diffusing capacity or both, related to interstitial or pleural changes on chest radiography. Newball et al found evidence of airway disease in six out of 13 patients evaluated. They concluded that certain patients with Sjögren's syndrome develop an unusual type of chronic obstructive airway disease which is probably the result of a chronic inflammatory process similar to that seen in their salivary and lacrimal glands. The authors suggested that the restrictive ventilatory defect seen in Sjögren's syndrome might be associated with the connective tissue disorder rather than with the sicca complex.

The purpose of this study is to report the types of pulmonary functional defects seen in 20 patients with Sjögren's syndrome.

Methods

Twenty non-smokers, 19 women and one man, who were diagnosed as suffering from Sjögren's syndrome or sicca complex, underwent clinical examination, chest radiography, and standard pulmonary function tests. Keratoconjunctivitis sicca was present in 19 out of 20 patients and included positive Shirmer test and punctate ulcers on cornea and conjunctiva found by slit lamp or fluorescent staining. Xerostomia was present in all patients and included diminution of salivary flow, positive sialography, and focal lymphoplasmacytic infiltrates and chronic fibrosing sialadenitis on lower lip or parotid biopsy. Connective tissue disease was present in seven patients (35%). Five patients met the criteria for the diagnosis of rheumatoid arthritis, one for dermatomyositis, and one for scleroderma.

Spirometry was performed with a Godart water spirometer. The FRC was determined with the helium dilution technique and thoracic gas volume (TGV) and airways resistance (Raw) were determined plethysmographically. Flow volume curves were determined by an Ohio spirometer and recorded on a Hewlett-Packard x-y recorder. Carbon monoxide diffusing capacity was determined by the steady state method. Arterial blood gases were measured in duplicate with standard electrodes (Radiometer, Model BMS-3).

Results

The anthropometric and clinical characteristics of the patients are listed in Table 1. The group consisted of 19 women and one man with a mean age...
of 65.7 years and age range of 34–79 years. The
diagnostic tests included Shirmer test, which was
positive in 19 out of 20 patients. Slit lamp exami-
nation revealed erosions on the corneal and con-
junctival epithelium in 13 out of 15 patients
examined. The two patients with negative slit lamp
examination had a positive Shirmer test. Mixed
salivary flow rate was decreased or absent in all
the patients. Lower lip (minor salivary gland) or
parotid biopsy revealed massive lymphoid infil-
tration with acinar atrophy and fibrosis in 14 out of
15 patients. Abnormalities related to narrowing of
the ducts and acinar atrophy were demonstrated on
sialography in six out of eight patients studied.

Chest radiographs were abnormal in seven
patients. The most common abnormality was
hyperinflation in four cases. Reticulonodular
pattern was demonstrated in one patient, who was
suffering from recurrent pulmonary infections.
Small lungs with “ground glass” appearance were
found in patients suffering from scleroderma.
Chest radiographs of the only male patient dem-
onstrated a diffuse interstitial infiltration. Trephine
biopsy of the right lung in this patient re-
vealed the pathological picture compatible with
lymphoid interstitial pneumonia (LIP).

The laboratory findings revealed anaemia with
haemoglobin less than 12 g/dl in six patients,
sedimentation rate in excess of 50 mm/1h in seven
patients. Rheumatoid factor was present in 15
patients (75%) and antinuclear factor in eight out of
18 studied. Increased levels of IgA were demon-
estrated in eight patients, while seven had increased
levels of IgM and four of IgG.

Pulmonary function tests were normal in eight

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<tr>
<th>Number</th>
<th>Sex</th>
<th>Age (yr)</th>
<th>Height (cm)</th>
<th>BSA</th>
<th>Associated immune disease</th>
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<th>Shirmer test</th>
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RA = rheumatoid arthritis; LIP = lymphoid interstitial pneumonia; + = positive test; − = negative test; No = test not performed.
patients (table 2). In seven patients pulmonary function tests demonstrated obstructive ventilatory defects. Most of the abnormalities were mild to moderate. Airway resistance was elevated and the specific airway conductance reduced in all cases. All but one patient had normal or high carbon monoxide transfer factor. Arterial blood gas analysis showed mild hypoxia with hypocapnia caused by alveolar hyperventilation in all patients. Increased alveolar-arterial (A-a) oxygen gradient on room air was observed in all seven patients.

A restrictive ventilatory defect was demonstrated in three patients. The pulmonary diffusing capacity was low in two patients and airway mechanics were normal in all three patients.

In two patients a mixed obstructive and restrictive ventilatory defect was found. There was a reduction in lung volumes with a low VC, FRC, and TLC and a reduction in the FEV1/FVC ratio with even more prominent reduction in the late flows. The specific airway and transfer factor were also reduced.

Discussion

The frequency of pulmonary involvement in Sjögren's syndrome is quite high. Sixty per cent of our patients had pulmonary functional abnormalities of some degree, a similar finding to that of Newball et al.\textsuperscript{4} The functional derangements in Sjögren's syndrome may be obstructive, restrictive, or mixed. The obstructive ventilatory pattern was the most common and was of mild to moderate severity. In this group in only one patient was the transfer factor diminished without physiological evidence of emphysema. Coury et al\textsuperscript{5} and Lemercier et al\textsuperscript{6} first reported on the occurrence of obstructive ventilatory defects of various severity in three out of five patients studied. Obstructive ventilatory defect was the only abnormality in six patients studied by Newball.\textsuperscript{4} The pathological basis for airway obstruction in Sjögren's syndrome involves lymphoid infiltrates and atrophy of mucous glands of pharynx, larynx and bronchial tree.\textsuperscript{3} The involvement of the lower respiratory tract by lymphocytic and plasma cell infiltrates around the walls of small airways gives rise to focal atelectasis, recurrent infections and eventually bronchiectasis.\textsuperscript{7-9}

A restrictive ventilatory defect was observed in only three of our patients with Sjögren's syndrome. All three had the complete syndrome. Strimlan et al\textsuperscript{3} found a restrictive ventilatory defect with or without decrease in the diffusing capacity in all 18 patients who underwent pulmonary function studies. It seems that restrictive defects are a common feature of Sjögren's syndrome.\textsuperscript{10-11} It is claimed that because the connective tissue disorders such as scleroderma and rheumatoid arthritis frequently have associated interstitial pulmonary fibrosis,\textsuperscript{8} the restrictive defect seen in the complete syndrome is more likely to be associated with the connective tissue disorder, rather than with the sicca complex per se. The spectrum of abnormalities leading to the restriction in Sjögren's syndrome includes also lymphoid interstitial pneumonia (LIP),\textsuperscript{14} pseudolymphoma,\textsuperscript{13-17} and malignant lymphoma of the lung.\textsuperscript{18-20}

A mixed type of pulmonary functional abnormality was observed in two patients, one of whom had the sicca complex and the other the complete variant. This type was the least common ventilatory abnormality seen in our patients and such cases could not be found in the English medical literature.

References

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