

Brief communications

Primary Sjögren's syndrome associated with recurrent sinopulmonary infections and bronchiectasis

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Sjögren's syndrome (SS) is an autoimmune disorder with unique involvement of the eyes and respiratory tract. Patients with SS frequently come to the allergist for evaluation.¹ We present the case of patient referred for evaluation of recurrent respiratory infections to rule out a diagnosis of immune deficiency.

CASE REPORT

A 58-year-old woman was evaluated for a history of recurrent sinopulmonary infections. Medical history revealed chronic rhinitis that was not associated with seasonal exacerbations. She had received antibiotics on many occasions for treatment of clinical sinusitis. She had four episodes of pneumonia associated with fever, productive cough, and an abnormal chest roentgenogram. She had noted the gradual onset of dry eyes and dry mouth. She noted intermittent episodes of painless swelling of the parotid glands. The sicca symptoms were not associated with medication use. Her pulmonary infections were easily treated with standard antibiotics, but between episodes she would have a dry, nonproductive cough. Results of system review were negative.

As determined by physical examination the patient had dry eyes, dry mouth, numerous repaired dental caries, and supple neck without lymphadenopathy, thyromegaly, or enlarged salivary glands. Her pulmonary examination was remarkable for persistent rales heard over the left lower lobe. No wheezing was heard. Her heart examination was unremarkable. Abdominal examination was unremarkable. No lymphadenopathy was noted. There were no rashes.

The results of complete blood cell count, chemistry profile, liver profile, urinalysis, thyroid profile, immunoglobulins, and complement studies were normal. The test results for, rapid plasmin reagin, human immunodeficiency virus, cryoglobulins, and cold agglutinins were negative. The sedimentation rate was 76. The protein electrophoresis showed increased α -2 and β globulin. The antibody to nuclear antigens was 2+,

Abbreviations used

CT: Computed tomography
SS: Sjögren's syndrome

with a speckled pattern. The rheumatoid factor, anti-DNA, anti-extractable nuclear antigen, and anti-histone were negative. The anti-SS-A (Ro) and anti-SS-B (La) were positive. Skin prick test responses to various inhalants, molds, and pollens were negative. Spirometry results were normal. Chest computed tomography (CT) showed pulmonary infiltrates of the left lower lobe and no adenopathy or masses. High-resolution chest CT showed bronchiectasis of the left lower and right middle lobes, associated with linear fibrotic changes. The tuberculin skin test result was negative, and the *Candida* control was positive.

The results of Schirmer's test, rose bengal test, and slit-lamp examination were consistent with keratoconjunctivitis sicca. A minor salivary gland biopsy was refused by the patient. A diagnosis of probable primary SS was made, and supportive measures were started.

DISCUSSION

The sicca complex clinically defines SS. Autoimmune exocrinopathy describes the progressive mononuclear inflammatory destruction of exocrine glands.¹ SS may have systemic manifestations in addition to the sicca complex. SS can occur alone (primary SS) or may be associated with another connective tissue disorder (secondary SS). Secondary SS is most commonly associated with rheumatoid arthritis, and less commonly, with other autoimmune syndromes. SS may be associated with benign or malignant lymphoproliferative disorders, and patients with SS have a 44-fold increased risk of developing lymphoma, compared with control subjects.¹

The major clinical manifestations of SS are keratoconjunctivitis sicca, xerostomia, and recurrent episodes of salivary gland enlargement.³ Other exocrine glands may be involved. Xerotrachia and xerobronchia may cause a dry, nonproductive cough, recurrent episodes of pneumonia, or bronchiectasis.² Other complications of the

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sicca complex include recurrent otitis media, chronic sinusitis, gingivitis, and dental caries. Systemic complications may be manifested as organ-specific autoimmune disorders.¹

There are no universally accepted criteria for the diagnosis of SS. The diagnosis is established by performing an ophthalmologic examination and a minor salivary gland biopsy. The ophthalmologic examination includes a Schirmer's test and a rose bengal dye test to confirm keratoconjunctivitis sicca. If autoantibodies (i.e., antibody to nuclear antigens, rheumatoid factors, anti-Ro, anti-La) are present, they add confirmation to the diagnosis of SS or related autoimmune diseases. The absence of autoantibodies, however, does not rule out the diagnosis of SS.

Roentgenographic diagnosis of lung complications in SS is greatly aided by CT. Plain chest CT images did not demonstrate lymphomatous changes or bronchiectasis in our patient. High-resolution CT images easily demonstrated bronchiectasis. High-resolution CT has been shown to be a sensitive tool for diagnosing bronchiectasis.⁴

Symptomatic relief and prevention of complica-

tions are important issues in treating SS. Medications or factors that exacerbate the drying of mucous membranes should be discontinued. Dry mucous membranes should be rehydrated. Infections are treated aggressively to prevent complications such as bronchiectasis. Patients with SS require close follow-up because of their propensity to develop lymphoproliferative disorders.^{1, 2, 5} High-resolution chest CT should be performed in patients who have pulmonary complications related to SS.

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***Aspergillus* brain abscess complicating allergic *Aspergillus* sinusitis**

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The ubiquitous fungus *Aspergillus* causes a wide spectrum of clinical syndromes, including allergic bronchopulmonary aspergillosis (ABPA)¹ and allergic *Aspergillus* sinusitis (AAS).² These latter syndromes reflect hypersensitivity responses to fungal antigens in the absence of fungal invasion. The majority of affected individuals are young, atopic, and immunocompetent. We describe a unique case of allergic *Aspergillus* paranasal pansi-

Abbreviations used

- AAS: *Aspergillus* sinusitis
ABPA: Allergic bronchopulmonary aspergillosis
CT: Computed tomography
FEV₁: Forced expiratory volume in 1 second
FVC: Forced vital capacity

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nitis in a patient who had an *Aspergillus* brain abscess and concomitant ABPA.

CASE REPORT

A 32-year-old Indian man with a history of asthma and chronic nasal obstruction who had undergone bilateral Caldwell-Luc sinus drainage procedures in 1986, came to the emergency department in March 1990 after