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Case report

Primary Sjögren's Syndrome Associated with Non-Hodgkin's Lymphoma of Salivary Gland and Cystic Lung Disease

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A B S T R A C T

A rare case of a young nonsmoker woman with Sjögren's syndrome and salivary gland non-Hodgkin's lymphoma, diagnosed one year later, is presented. Three years after treatment of the lymphoma, asymptomatic progression of the Sjögren's syndrome was observed with pulmonary involvement – predominantly bullous or cystic lung disease.

To our knowledge, this is the only report of Sjögren's syndrome associated with non-Hodgkin's lymphoma in salivary gland, and complicated with multiple lung cysts.

Key words: *Sjögren's syndrome, non-Hodgkin's lymphoma, salivary gland, lymphocytic interstitial pneumonia, cystic lung disease*

Introduction

Sjögren's syndrome is a chronic inflammatory autoimmune disorder characterized by lymphocytic infiltration of the exocrine glands. The diagnostic criteria include keratoconjunctivitis sicca and xerostomia. Additionally, different types of lung involvement have been described in Sjögren's syndrome of which the bullous/cystic lung disease is a very rare manifestation[1,2].

Patients with Sjögren's syndrome are at increased risk of lymphoma development. Most of these lymphomas involve the neck organs[3]. Salivary gland mucosa-associated lymphoid tissue (MALT) lymphomas are very rare, but their close association with autoimmune diseases confirms their possible role in pathogenesis of these lymphomas[4].

We describe a case of Sjögren's syndrome associated with non-Hodgkin's lymphoma in salivary gland, and complicated with multiple lung cysts.

Case report

In nonsmoker female patient, born in 1973, the diagnoses of Sjögren's syndrome and sialolithiasis of the left parotid gland were confirmed in 1999. The diagnosis of Sjögren's syndrome was based on classification criteria by Vittali et al.[5] with patient's positive ocular and oral symptoms, minor salivary gland biopsy, diagnostic test for salivary gland involvement and raised levels of auto antibodies. The patient reported enlarged neck lymph nodes in October 2000 followed by extirpation of an enlarged lymph node and left submandibular gland. Histology confirmed marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT) type in salivary gland and lymph node follicular hyperplasia. She had been received chemotherapy till June 2001 and radiotherapy of salivary glands region afterwards.

The initial chest radiogram was normal as well as follow-ups every 6 months until April 2004 when rare discrete parenchymal opacities measured 1 – 2 cm have been depicted in both lower lung zones. High-resolution computed tomography (HRCT) scans showed multiple cystic formations predominantly in the middle and lower lung zones (Figure 1), measured from 1 to 4 cm in diameter. Four parenchymal consolidations were found in the same areas, measured from 0.5 to 2 cm (Figure 2). The findings were interpreted as pulmonary cysts of unknown origin (not resembling those in lymphangioleiomyomatosis, Langerhans cell histiocytosis or irregular emphysema) and parenchymal consolidations suggesting infectious origin with Pneumocystis carinii as a possible agent.

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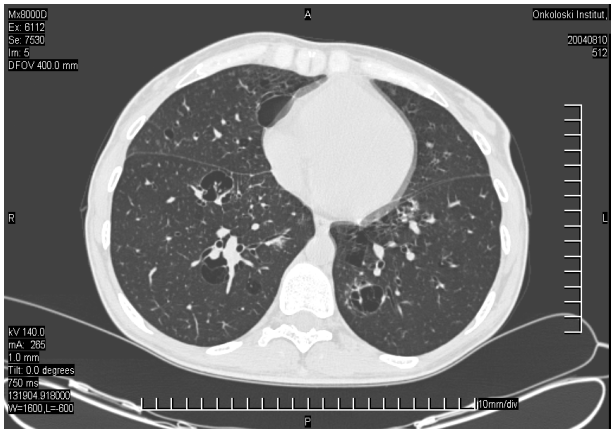

Fig. 1. High-resolution computed tomography in August 2004: Multiple lung cystic lesions.

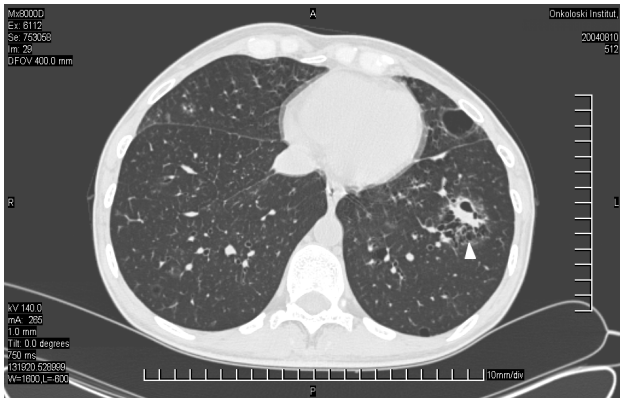

Fig. 3. High-resolution computed tomography in August 2004 at the same level as Figure 2. The most of parenchymal consolidation was replaced by small thin walled cysts.

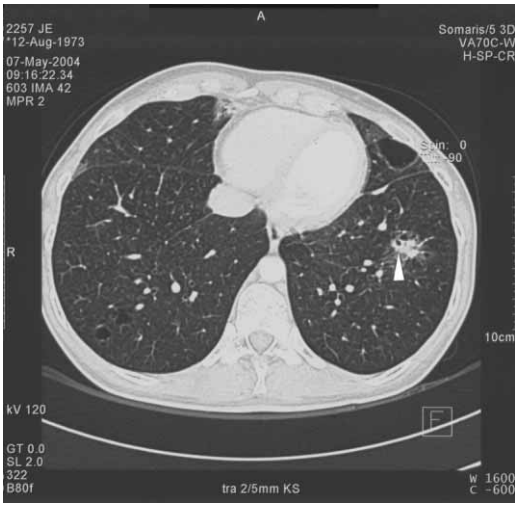

Fig. 2. High-resolution computed tomography in May 2004: Parenchymal consolidation in the left lower lobe.

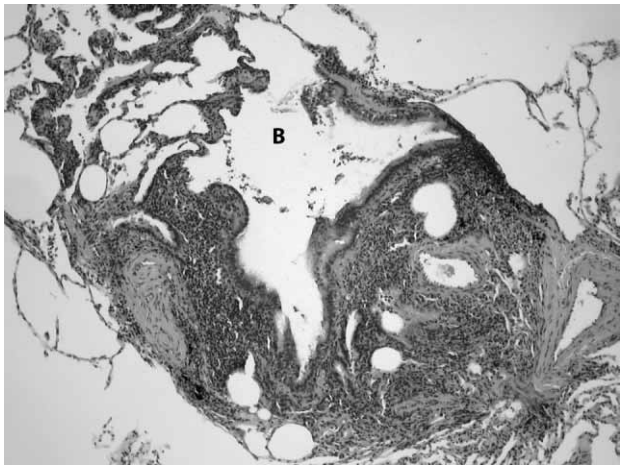

Fig. 4. Transbronchial biopsy: cystic-like bronchioloectasia (B) with lymphocytic infiltration in bronchiolar walls.

The patient had no respiratory symptoms although pulmonary function tests revealed mild restrictive ventilatory disfunction (VC=3.7 *l*, %VC=80%, FEV_1=3.4 *l,* FEV_1 % =92%, $\%DL_{CO}$=64%). Transbronchial biopsy specimen was interpreted as chronic non-specific inflammation with no signs of lymphoma infiltration. *Streptococcus pneumoniae* and *Candida glabrata* were isolated from the alveolar lavage and the patient was consecutively treated by antimycotic drugs for four weeks without regression of parenchymal consolidations on chest radiographs.

The follow-up HRCT showed progression of pulmonary cystic lesions in number and size (Figure 1). Some of the parenchymal consolidations were transformed into small cysts (Figure 3), suggesting a rare cystic/bullous form of Sjögren's syndrome in the lung.

The second transbronchial biopsy taken from left lower lobe consolidation showed irregular and intense lymphocytic interstitial infiltration with widened small airways (Figure 4). Alveolar lavage taken from this area was interpreted as CD8 lymphocytic alveolitis with CD4/CD8 index of 0.5, thus confirming autoimmune involvement of pulmonary interstitium.

Discussion

Sjögren's syndrome is an autoimmune disease with lymphocytic infiltration of glandular and extraglandular tissues[6]. In most patients, it is confined to the salivary and lacrimal glands, however, extraglandular infiltration has been identified in 5 to 10% of the affected patients[7].

The lung with abundant mucosal glands is a primary target. Several pulmonary complications have been described, including interstitial lung disease, airway disease, and nodular infiltrates[8]. Cystic or bullous disease is a rare complication of Sjögren's syndrome with only 15 cases in the literature[1,2]. Compared to the other cases, our patient is the youngest of all with cystic lung disease.

Meyer et al[9] suggested that stenosis with obstruction of bronchioles by peribronchiolar lymphocyte infiltration is the cause of air trapping with check-valve mechanism leading to formation of the multiple lung cysts. On the contrary, Johkoh et al[10] proposed lymphocytic interstitial pneumonia to be the common feature in the cases of thin-walled cystic formations. Nevertheless, the radiological differential diagnosis of lung cysts in Sjögren's syndrome includes cystic abnormalities associated with Langerhans cell histiocytosis, lymphangioleiomyomatosis, different types of emphysema, honeycomb cysts in idiopathic pulmonary fibrosis, and thin walled cysts in *Pneumocystis carinii* pneumonia.

We are presenting the first report of Sjögren's syndrome associated with non-Hodgkin's lymphoma in salivary gland and complicated with multiple lung cysts and some parenchymal consolidations. Therefore one should know that such lung manifestations of Sjögren's syndrome could be also associated with salivary gland MALT lymphoma.

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PRIMARNI SJÖGRENOV SINDROM UDRUŽEN S NE-HODKINOVIM LIMFOMOM PAROTIDNIH ŽLIJEZDA I CISTIČKOM BOLEŠĆU PLUĆA

SAŽETAK

Prikazan je vrlo rijedak slučaj mlađe žene, nepušačice, s Sjögrenovim sindromom, koja je godinu dana nakon postavljanja diagnoze oboljela također i za ne-Hodkinovim limfomom parotidnih žlijezda. Nakon tri godine liječenja limfoma pojavljuje se asimptomatsko napredovanje Sjögrenove bolesti u plućima sa buloznim ili cističkim promjenama pluća. Prema našim podatcima radi se o prvom prikazu Sjögrenovog sindroma s ne-Hodkinovim limfomom parotidnih žlijezda, koji se zakomplicirao cističkom bolešću pluća.