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## Primary Sjögren's syndrome with two extraglandular sites involvement — case report

Zespół Sjögrena z zajęciem dwóch narządów pozawydzielniczych — opis przypadku

### Abstract

Primary Sjögren's Syndrome (pSS) is a chronic, slowly progressive inflammatory autoimmune disorder, characterised by lymphocytic infiltration of the exocrine glands, leading to decrease of glandular secretion. In 40–60% of pSS patients, extraglandular disease develops.

We present the case of a patient with two extraglandular sites involvement in the course of pSS manifesting with progressive respiratory and gastrointestinal symptoms.

**Key words:** primary Sjögren's syndrome, diagnosis, lung disease, gastrointestinal disease

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### Streszczenie

Pierwotny zespół Sjögrena (pSS) jest przewlekłą postępującą chorobą zapalną o podłożu autoimmunologicznym, charakteryzującą się naciekiem limfocytarnym gruczołów wydzielania zewnętrznego, który prowadzi do zmniejszenia funkcji wydzielniczej tych gruczołów i w konsekwencji — do zespołu suchości. U 40–60% chorych dochodzi do rozwoju pozagruczołowych objawów choroby.

W pracy opisano przypadek chorej na pierwotny zespół Sjögrena, przebiegający z zajęciem płuc i układu pokarmowego.

**Słowa kluczowe:** pierwotny zespół Sjögrena, rozpoznawanie, choroby płuc, choroby układu pokarmowego

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### Introduction

Sjögren's syndrome (SS) is a chronic, slowly progressive, inflammatory autoimmune disorder, characterised by lymphocytic infiltration of the

exocrine glands, leading to decrease of glandular secretion [1]. The disease may occur alone (primary SS) or in association with other autoimmune diseases such as rheumatoid arthritis, systemic lupus erythematoses and scleroderma (secondary SS)

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[2–4]. The prevalence of primary SS (pSS) is 0.5–1% in the general population [1, 3]. Most patients with pSS present only with the symptoms of keratoconjunctivitis sicca and xerostomia. In 40–60% of patients, extraglandular disease develops [2], which may be manifested by:

- lymphoepithelial lesions in extra-exocrine gland tissues (bronchi, renal tubules, biliary ducts)
- vasculitis related to the deposition of immune complexes due to B-cell hyperreactivity.

Sicca syndrome may also be observed in the course of sarcoidosis, viral infections (HCV, HIV), and as a result of radiotherapy of the head and neck [1, 3].

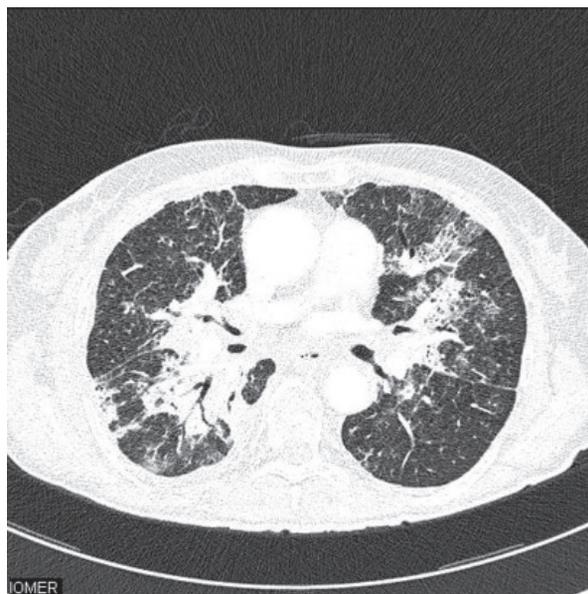
We present the case of a patient with two extraglandular sites involvement in the course of primary SS, manifesting with progressive respiratory and gastrointestinal symptoms.

### Case report

A 71 year-old Caucasian woman, an agricultural worker, with a history of arterial hypertension and a healed gastric ulcer, was referred to the Department of Chest Medicine of our Institute in March 2005 due to fatigue, dyspnea on exertion, chest tightness, epigastric pain and progressive weight loss (16 kg in eight months). In 2002, primary Sjögren's syndrome had been diagnosed based on: keratoconjunctivitis sicca with positive Schirmer test and positive result of minor salivary gland biopsy.

On admission, she was in good status, slim, but not cachectic. Physical examination revealed sicca syndrome and crepitations over the base of both lungs.

Blood morphology and serum biochemical parameters were within normal limits. Total protein count was 7.1 g/L, with 26% of gamma globulin. ESR — 70/hour, CRP — 21 mg/L, ANA 1:1280 of homogenous pattern. Type of antinuclear antibodies was not defined (Ro/SS-A and La/SS-B — were not found). Chest X-ray examination revealed bilateral interstitial infiltrates. Chest CT scan contained areas of consolidations, with peribronchial and subpleural distribution, some of them with air-bronchogram. Focal ground-glass attenuation areas were also seen, as well as reticular and linear opacities with traction bronchiectases localised in lower lung fields. Mediastinal lymph nodes were moderately enlarged (aorto-pulmonary — 16 mm, subcarineal — 20 mm in diameter, with calcifications). The morphology of lung attenuations seen on HRCT suggested, in the opinion of the radiologist, the presence of non-specific interstitial pneumonia (NSIP) and organising pneumonia (OP) (Fig. 1).



**Figure 1.** Chest CT scan (April 2005) — areas of consolidations, with peribronchial and subpleural distribution, some of them with air-bronchogram. Focal ground-glass attenuation areas, reticular and linear opacities with traction bronchiectases localised in lower lung fields

**Rycina 1.** Badanie TK klatki piersiowej (kwiecień 2005) — obszary zagęszczeń zlokalizowane okołooskrzelowo i podopłucnowo, część z nich zawiera bronchogram powietrzny. Ogniska matowej szyby, zmiany siateczkowate i linijne z rozstrzeniami oskrzeli z pociągania w dolnych częściach płuc

The results of arterialised capillary blood gas analysis were: PaO<sub>2</sub> — 60.9 mm Hg, PaCO<sub>2</sub> — 33.8 mm Hg, pH — 7.45. In the six-minute walking test (6MWT) she covered 366 meters with desaturation (92–87%). Systolic pulmonary artery pressure (PASP) estimated with echocardiography was 41 mm Hg, suggesting the possibility of pulmonary hypertension. Pulmonary function tests (PFT) revealed normal volumetric measures, but markedly decreased static compliance — 0.76 L/kPa (29% of predicted) and diffusion capacity for carbon monoxide (DLCO) — 2.4 mmol/min/kPa (39% of predicted).

Bronchoscopy identified a chronic inflammatory process in the bronchial tree. Bronchoalveolar lavage (BAL) counts included: 66% of macrophages, 15% of polymorphonuclear granulocytes, 18% of lymphocytes, CD4/CD8 — 1.37. Transbronchial lung biopsy was performed. The histological examination revealed the presence of granulomas formed of epithelioid and large cells, without necrosis. The pathologist's diagnosis was sarcoidosis or other granulomatous lung disease. Results of cultures of bronchial secretions and tissue staining for mycobacteria and fungi were negative.

Gastroscopy was also performed and extensive gastritis with thickened gastric rugae was found. The histological examination of gastric mucosa revealed the presence of lymphocytic infiltration, highly suspicious of lymphoma. The specimens were examined by a haematologist-pathologist and the final diagnosis was chronic active gastritis with the signs of destruction of epithelium lining mucosal glands, possibly in the course of Sjögren's disease.

To make a final diagnosis of lung pathology, right antero-lateral thoracotomy with open lung biopsy was performed. Lung was of high density with palpable nodules 2–10 mm in size; the specimens were taken from right lower lobe.

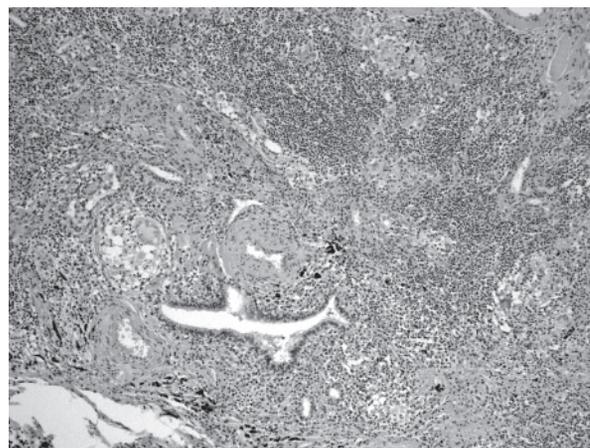
On the fifth post-operative day, the patient deteriorated, with increased body temperature, pronounced dyspnea and strong abdominal pain localised in the right subcostal region. Chest X-ray revealed progression of lung infiltrates. Ultrasonographic examination of abdominal cavity revealed the presence of enlarged gall bladder with hypoechogenic halo; acute acalculous cholecystitis was diagnosed. The patient received symptomatic treatment with broad spectrum antibiotics, spasmolytics, parenteral nutrition and oxygen therapy. Abdominal symptoms decreased, but respiratory symptoms persisted. The results of bacteriological cultures of blood and sputum were negative. Progression of the immunological process involving lungs and gastrointestinal system was considered as one of the possible causes of deterioration. Thus, the patient received pulses of solumedrol (500 mg intravenously on four consecutive days). Marked clinical improvement and regression of lung infiltrates was noted on the 14th post-operative day. At that time, the results of open lung biopsy were obtained.

The microscopic examination of lung tissue demonstrated diffuse, highly cellular infiltrate composed of small mature lymphocytes and plasma cells with few reactive follicles (Fig. 2).

Numerous, small and non-necrotising granulomas containing multinucleated giant cells and epithelioid cells were identified in the interstitium of the lung.

Lymphoid infiltrates were distributed along the bronchovascular bundles and were seen within intra-alveolar septa expanding them. Scattered larger lymphoid cells were noticed on the edge of the infiltration. Occasionally, lymphoid infiltrates of bronchial, bronchiolar epithelium were also seen (so called 'lymphoepithelial lesions'). Focal interstitial fibrosis with destruction of lung parenchyma was found.

Immunohistochemical studies showed that the cellular infiltrate that distended the alveolar



**Figure 2.** Sjögren's syndrome. Fragment of pulmonary tissue with diffuse lymphoid infiltrate and interstitial fibrosis. Poorly formed granulomas with multinucleated giant cells seen.

Microphotograph, stain H+E, high magnification

**Rycina 2.** Zespół Sjögrena. Fragmenty tkanki płucnej z rozlanym naciekiem z komórek limfoidalnych i włóknieniem śródmiąższowym. Słabo uformowane ziarniniaki zawierające komórki olbrzymie

walls was composed primarily of T cells, with relatively few intermingled B lymphocytes and with a low proliferation index. Furthermore, some special histochemical and immunohistochemical stains were performed to exclude lung infection.

The lymph node of group 7 taken during open lung biopsy contained hyalinised collagen stroma with numerous dust-laden macrophages, without granulomas. On the basis of the microscopic picture and immunohistochemical reactions, a diagnosis of lymphocytic interstitial pneumonia (LIP) in the course of pSS was established.

Thus the final clinical diagnosis was primary Sjögren's syndrome with progressive lung and gastrointestinal system involvement. The patient was further treated with prednisolone 1 mg/kg/day and cyclophosphamide in pulses (100 mg/day per os for ten days each month). After eight months of such treatment, the patient was well, with improved exercise tolerance and no abdominal complaints. During 6MWT, she covered 384 metres, without desaturation. The control HRCT scan revealed marked regression of lung parenchymal infiltrates (Fig. 3). Lung volumes remained within normal limits, DLCO was still low: 2.52 mmol/min/kPA (42% of predicted). On endoscopic examination, gastric mucosa looked normal and histological examination revealed no abnormalities. The severe adverse effects of the steroidotherapy carried out in 2006 (osteoporosis, diabetes mellitus and glaucoma) were observed. The patient was advised to decrease gradually the dose of steroids up to 5 mg/day.



**Figure 3.** CT scan (December 2005) — marked regression of pulmonary consolidations after course of treatment

**Rycina 3.** Badanie TK klatki piersiowej (grudzień 2005) — znacząca regresja obszarów zagęszczeń po leczeniu

Cyclophosphamide was also reduced to 100 mg/day for seven days in a month, and in March 2007 it was stopped. In December 2007, clinical deterioration was diagnosed: the patient complained of poorer exercise tolerance, dry cough and arthralgia. ESR was 80/hour, CRP — 15 mg/L, ANA 1:5120, RF — negative. Distance of 6MWT had decreased to 266 metres, with desaturation (95–91%). Chest HRCT scan revealed no new pathological changes; the results of echocardiography revealed no progression of pulmonary hypertension. Azathioprine was introduced (100 mg/day) with subsequent clinical improvement. On the last visit (March 2010) the patient was well, with no signs of disease progression.

## Discussion

The diagnosis of primary Sjögren's syndrome in the presented patient was based on: ocular symptoms of inadequate tear production with signs of corneal damage and positive Schirmer test, oral symptoms of decreased saliva production and the results of minor salivary gland biopsy demonstrating lymphocytic infiltration. Despite the presence of serum autoantibodies in high titre, anti SS-A/Ro and anti SS-B/La were not found. Nevertheless, the patient's disease fulfilled diagnostic criteria of pSS according to American-European classification [5] (Tab. 1). According to data from

the literature, 10–40% of pSS patients lack Ro or La antibodies in serum, despite the presence of ANA in high titre [6–8].

The symptoms of lung and gastrointestinal system involvement developed in our patient three years after initial diagnosis. The reported incidence of lung involvement in the course of pSS varies widely from 9 to 75%, partly due to the lack of universal agreement over the diagnostic criteria of the disease [4, 9]. Hatron et al. performed BAL in asymptomatic pSS patients and found lymphocytic alveolitis in 55% of them [10]. BAL performed in our patient contained moderately increased number of T lymphocytes and polymorphonuclears.

The radiological manifestation of lung disease in the presented case consisted of bilateral areas of consolidations with peribronchovascular localization, focal ground glass opacities and traction bronchiectases. Moderately enlarged mediastinal lymph nodes were also described. The pattern of lung consolidations was described as suggestive for NSIP coexisting with OP.

HRCT abnormalities in pSS with lung involvement have been found by several authors [11–13]. Both parenchymal pathology and small airways disease have been described in the literature. Linear and reticular opacities with peribronchovascular distribution [4, 6, 14], areas of ground-glass attenuation [15] as well as traction bronchiectases and septal thickening [14] localised mostly in the lower lobes have been the most frequent findings. The presence of ground-glass opacities and thin wall cysts are indicative of lymphocytic interstitial pneumonia (LIP) [11, 16], but the correlation between the radiological and the histological picture in other types of lung pathology is poor [6]. Thus lung biopsy is often performed to establish the correct diagnosis [17].

Specimens obtained in our patient from transbronchial biopsy revealed the presence of noncaseating sarcoid-like granulomas. Histopathological examination of the material obtained from open lung biopsy demonstrated extensive lymphoid infiltrate within interalveolar septa. Sarcoid-like granulomas were present in the interstitium, but not in dissected lymph node. Thus sarcoidosis was ruled out as the cause of lung disease, and the final diagnosis was LIP.

Various histological patterns of interstitial lung diseases such as non-specific interstitial pneumonia (NSIP), usual interstitial pneumonia (UIP), LIP, organising pneumonia (OP), primary pulmonary lymphoma and diffuse interstitial amyloidosis have been reported in pSS patients [2, 4, 18–20]. The predominance of LIP was noted in earlier studies, but recen-

**Table 1. Revised international consensus criteria for Sjögren's syndrome [5]****Tabela 1. Międzynarodowe kryteria klasyfikacyjne zespołu Sjögrena [5]****I. Ocular symptoms (at least one present)**

1. Persistent, troublesome dry eyes every day for more than three months
2. Recurrent sensation of sand or gravel in the eyes
3. Use of a tear substitute more than three times a day

**II. Oral symptoms (at least one present)**

1. Feeling of dry mouth every day for at least three months
2. Recurrent or persistent feeling of swollen salivary glands as an adult
3. Need to drink liquids when swallowing dry food

**III. Ocular signs — objective evidence of ocular involvement (at least one present)**

1. Schirmer's I test — performed without anesthesia ( $\leq 5$  mm in five minutes)
2. Rose-Bengal score or other ocular dye score ( $\geq 4$  according to van Bijsterveld's scoring system)

**IV. Histopathology**

In minor salivary glands (obtained through normal appearing mucosa) focal lymphocytic sialadenitis, evaluated by an expert histopathologist, with a focus score  $\geq 1$ , defined as a number of lymphocytic foci (which are adjacent to normal-appearing mucous acini and contain more than 50 lymphocytes) per 4 mm<sup>2</sup> of glandular tissue

**V. Objective evidence of salivary-gland involvement (at least one present)**

1. Unstimulated whole salivary flow ( $\leq 1,5$  ml in 15 min)
2. Parotid sialography showing the presence of diffuse sialectasias (punctuate, cavitory or destructive pattern), without evidence of obstruction in major ducts
3. Salivary scintigraphy showing delayed uptake, reduced concentration and/or delayed excretion of tracer

**VI. Autoantibodies: presence in the serum of the following autoantibodies**

Antibodies to Ro (SS-A) or La (SS-B), or both

Primary SS (in patient without any potentially associated disease):

— any four of six items as long as either IV (Histopathology) or VI (serology) is positive or three of four objective criteria items (III–VI)

tly NSIP and OP have been the most frequently reported types of lung pathology [2, 4, 8, 11].

Sarcoid-like granulomas have also occasionally been found in pSS patients [6, 21]. Authors have disagreed whether these cases represented pSS coexisting with sarcoidosis [22–24] or sarcoidosis mimicking pSS [25]. Recently, sarcoid-like granulomas have been described by pathologists as one of the possible compounds of cell infiltrate in the course of pSS [26, 27]. Nevertheless, in such circumstances, the possibility of sarcoidosis must be always ruled out, according to the latest American-European diagnostic criteria [5].

The other localisation of disease in our patient was the digestive system. Gastroscopy revealed the presence of hyperthrophic gastritis, and histological examination demonstrated lymphoid infiltration of gastric mucosa. The results of immunohistochemistry excluded gastric lymphoma and confirmed gastric infiltration in the course of pSS. Gastrointestinal involvement in pSS is common, mostly described as atrophic superficial gastritis. Dai et al. [28] reported a case of pSS with hypertrophic gastritis and biopsy proven lymphocytic

infiltration. Acute acalculous cholecystitis, recognised in the presented patient in the course of the disease, could also be a sign of gastrointestinal involvement in the course of pSS. The only such case was reported by Shin et al. [29] in a patient with SS secondary to lupus erythematoses. Nevertheless, Kochina et al. [30] found chronic cholecystitis in 51% of patients with Sjögren's syndrome.

Both pulmonary and gastrointestinal pathology observed in our patient required pathological examination to exclude malignancy. The overall prevalence of non-Hodgkin's lymphoma in pSS is ~5% [31–34]. The most frequent type is extranodal marginal zone lymphoma of the mucosa associated lymphoid tissue (MALT) [31–34]. Salivary glands are the most commonly affected site, but other extranodal sites such as the stomach, nasopharynx, skin, liver, kidney and lung can also be involved [31, 34].

The treatment of our patient consisted of corticosteroids in combination with cyclophosphamide. Most pSS patients with extraglandular disease receive corticosteroids alone [4]. The reason for introducing a cytotoxic drug was the involvement of the two extraglandular sites (lungs and stomach),

with severe respiratory insufficiency and acute acalculous cholecystitis in the course of the disease. The response to this drug combination was very good. Nevertheless, progression of symptoms was seen after cyclophosphamide was stopped. The second line therapy, with azathioprine, produced long-lasting clinical remission.

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