



## Sarcoidosis Presenting as Primary Sjogren's Syndrome

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Sarcoidosis is a chronic multi-system granulomatous disease of unknown etiology. Its most frequent manifestations include pulmonary involvement, fever, lymphadenopathy, skin lesions, splenomegaly, and musculoskeletal and eye involvement [1]. It can coexist with various connective tissue diseases including rheumatoid arthritis, systemic lupus erythematosus, systemic sclerosis and spondyloarthropathies [2]. Primary Sjogren's syndrome is a chronic autoimmune disease presenting with dry mouth and dry eyes and is characterized by a lymphoplasmocytic infiltrate involving the exocrine glands. The exocrine glands are rarely involved in sarcoidosis and most patients present with symptoms related to the respiratory system. The presentation of sarcoidosis with dry mouth and dry eyes is rare, challenging the clinician with the difficult task of differentiating SS from sarcoidosis. We describe a patient who presented with a one year history of dry mouth, dry eyes and parotid swelling but without respiratory symptoms, who subsequently was found to have sarcoidosis.

### Case Description

A 45 year old woman presented to the Sjogren syndrome clinic with a one year history of dry mouth and dry eyes. Three

months before her initial visit, bilateral parotid swelling was noted and the patient was seen by an otolaryngologist. Fine needle aspiration of the gland revealed scant lymphocytes suggestive of SS, and the patient was referred to the clinic for further evaluation.

Physical examination was normal except for mild bilateral parotid swelling and severe dryness of oral mucosa. Schirmers I-test was positive (2 mm5/min) and the parotid flow rate was significantly reduced (0.3 ml/min). Laboratory evaluation was unremarkable except for a positive antinuclear antibody test in titer of 1:160, with a speckled pattern and a rich growth of *Candida albicans* from a tongue culture. Biopsy of the minor salivary gland demonstrated the presence of non-caseating granulomas with scattered lymphoplasmocytic infiltrates. Specific stains for various microorganisms were negative. A chest X-ray following the biopsy showed bilateral hilar lymphadenopathy, and computed tomography scan revealed the presence of hilar adenopathy with minimal interstitial involvement. Pulmonary function tests, including CO diffusion, were within normal limits and serum angiotensin-converting enzyme levels were significantly elevated. Bronchoalveolar lavage yielded lymphocytosis of 25% with an H/S ratio of 4:7, and a transbronchial biopsy confirmed the diagnosis of sarcoidosis.

The patient was started on prednisone 10 mg/day, and within 2 weeks of treatment the symptoms of dry eyes and dry mouth had disappeared. The Schirmer test showed normal findings and parotid flow rates returned to normal. The patient is currently treated with prednisone 2.5 mg/day.

### Comment

Primary Sjogren's syndrome is a chronic autoimmune disease affecting mainly the exocrine glands, with the typical presentation of symptoms and signs related to dry mouth and dry eyes. On the other hand, sarcoidosis, which is a chronic systemic disease, is either asymptomatic or presents with respiratory symptoms such as dyspnea and dry cough, while symptoms of dry mouth or dry eyes are extremely rare [1]. Already in the 1960s a relationship between sarcoidosis and various autoimmune diseases was suggested. While initially considered rare, the coexistence of these disorders has lately been reported with increasing frequency [2]. The coexistence of sarcoidosis and SS is less frequent and has been reported in only 17 cases, whereas cases of autoimmune diseases complicating long-standing sarcoidosis have rarely been reported.

A review of the literature revealed 13 reported cases in which sarcoidosis presented as SS [Table 1]. These patients presented with xerostomia and xer-

SS = Sjogren's syndrome

**Table 1.** Sarcoidosis presenting as primary Sjogren's syndrome

Author	Age/Sex	Symptomatology	RF	ANA	Anti- Ro/La	Chest X-ray	ACE	Biopsy site
Melson et al. [4]	57/F	DE, PGE Weight loss	Neg	Neg	Neg	Normal	Increased	MSGB, LN
Drosos et al. [3]	47/F	DE, DM	Neg	1:160	Neg	Normal	NR	MSGB, TBLB
	50/F	DE, DM cough	Neg	1:160	Neg	HL	NR	MSGB
	60/F	DM, DE cough	Neg	1:320	Neg	ILD	NR	MSGB, TBLB
	64/M	DE, Dyspnea	Neg	1:80	Neg	ILD	NR	MSGB, TBLB
	44/F	DM	Neg	Neg	Neg	HL	NR	MSGB, TBLB
	41/F	DE, DM	Neg	1:160	Neg	NR	NR	MSGB
	50/F	DE, DM, PGE	Neg	Neg	Neg	HL	NR	MSGB, TBLB
	40/F	DE, DM cough	Neg	Neg	Neg	ILF	NR	MSGB, TBLB
	42/M	DE, DM, PGE	Neg	1:160	Neg	HL	NR	MSGB, TBLB
	38/F	DE, DM, PGE	Neg	1:80	Neg	HL	NR	MSGB
	35/F	DM, cough	Neg	Neg	Neg	ILD	NR	MSGB, TBLB
	46/F	DE, DM, PGE	Neg	Neg	Neg	HL	NR	MSGB, TBLB
	Present case	45/F	DE, DM, PGE	Neg	1:160	Neg	HL, ILD	Increased

ACE = angiotensin-converting enzyme, ANA = anti-nuclear antibodies, DE = dry eyes, DM = dry mouth, HL = hilar lymphadenopathy, ILD = interstitial lung disease, MSGB = minor salivary gland biopsy, NR = not reported, PGE = parotid gland enlargement, RF = rheumatoid factor, TBLB = transbronchial lung biopsy.

ophthalmia, but also with signs and symptoms of pulmonary or systemic disease such as cough, fever, fatigue and skin involvement [3]. Some of these patients presented with parotid gland enlargement, which was shown to be a useful clinical finding, since patients with sicca symptoms and parotid enlargement were more likely to have sarcoidosis than SS. Our case is unique in this respect since our patient suffered for more than a year from xerostomia, xerophthalmia and parotid enlargement without any symptom that could suggest pulmonary involvement or any sign of a systemic disease. Another interesting aspect of our case was the good response to treatment with low dose steroids, which would not be expected in SS alone. Such a response has not been reported previously; in fact, in none of the cases of sarcoidosis and SS that we reviewed was treatment of xerostomia and xerophthalmia mentioned at all. The fine needle aspiration of the parotid gland, which yielded only mature lymphocytes, further added to the diagnostic pitfall of SS. This diagnostic dilemma was also

reported by Melson et al. [4]: in their patient a lip biopsy showed histological features of SS.

The lip biopsy is an important diagnostic tool in SS, with high sensitivity and specificity, although according to the recently approved EC criteria for the diagnosis of SS this is not a prerequisite. It has been suggested that the lip biopsy is most necessary in patients who have partial San Diego criteria for SS and positive anti-Ro or anti-La antibodies [5]. In these patients the anti-Ro and/or anti-La positive antibodies will predict a positive biopsy in 85% of the cases and probable SS can be diagnosed. On the other hand, patients with partial San Diego criteria for sicca and negative anti-Ro and anti-La antibodies, like our patient, have a 82% chance for a negative biopsy, and the diagnosis of primary SS is nearly excluded [5]. The lip biopsy has also been used for the diagnosis of sarcoidosis, yielding typical non-caseating granulomas in about 50% of cases. Thus, our case further emphasizes the role of lip biopsy and the role of anti-Ro and anti-La serology in differ-

entiating between SS and a long list of diseases, including sarcoidosis, which can mimic the sicca syndrome.

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*Now this is not the end. It is not even the beginning of the end. But it is, perhaps, the end of the beginning*

*Sir Winston Churchill, British Prime Minister (1874-1965), on the Battle of Egypt, 1942*