

RESEARCH LETTER

Mortality of Bulgarian patients with primary and secondary Sjögren's syndrome

Panchovska M, Sheitanov Y, Uzunov N

Clinic of Rheumatology, Medical University, Plovdiv, Bulgaria. panchovska@abv.bg

Sjögren's syndrome is an autoimmune disease of exocrine glands, with the most common clinical presentations being xerostomia and xerophthalmia. The disease may manifest as primary Sjögren's syndrome (pSS) or associate with other rheumatic diseases – secondary Sjögren's syndrome (sSS) (1).

During the period March 1995 to May 2003 one hundred and seven patients with Sjögren's syndrome (European criteria, 1993) were followed.

There were sixty two patients with pSS, 45 patients with sSS (11 with rheumatoid arthritis, 3 with sclerodermia and 1 patient with systemic lupus erythematosus). The mean age of patients with pSS was 49.9 and of those with sSS 54.8 years. The frequency an causes of death were analysed according to the autopsy protocols and the hospital notesire medical records.

The results of the research established 18 death cases – 7 patients (11.29 %) with pSS and 11 patients with sSS (24.44 %). The frequency and causes of death in these two groups are presented in Table 1.

Tab. 1. Causes of death in patients with pSS and sSS.

Causes of death	Death cases (n)	
	pSS=7	sSS=11
lung disease*	-	2
pulmonary embolism**	-	1
acute respiratory failure	1	1
cardiac disease	-	1
cerebrovascular disease	-	2
sepsis	1	2
GI bleeding***	1	-
non lymphoma neoplasm	1	1
lymphoma	3	1

*connected with the primary disease, ** antiphospholipid syndrome,

*** stomach ulcer

The analysis of the death cases showed significantly higher mortality in patients with sSS than those with pSS.

Among the frequent causes of death are the pulmonary diseases, sepsis and brain vascular disease. The highest mortality is observed in patients with rheumatoid arthritis (n=7). Three death cases were registered in pSS with NonHodgkin Lymphoma (4.83 %). These data support the thesis for increased frequency of malignant lymphoproliferation in SS (4).

The results of the study show higher mortality in the sSS (especially rheumatoid arthritis), which is proven also in other studies (2). Probably there is a connection with the stage and the treatment of the primary disease, the accompanying diseases and side effects of medications.

Reference

- Manthorpe R.** New criteria for diagnosing Sjögren's syndrome: a step forward? *Scand J Rheum* 2001; 115 (Suppl): 14–22.
- Martens PB, Pillemer SR, Jacobsson LT et al.** Survivorship in a Population Based Cohort of Patients with Sjögren's Syndrome, 1976–1992. *J Rheumatol* 1999; 26 (6): 1296–1299.
- Vitali C, Bombardieri S, Moutsopoulos HM et al.** Preliminary criteria for the classification of Sjögren's syndrome. *Arthr Rheum* 1993; 36: 340–347.
- Voulgarelis M, Dafni UG, Isenberg DA et al.** Malignant lymphoma in primary Sjögren's syndrome. *Arth Rheum* 1999; 42: 1765–1771.

Received February 28, 2004.

Accepted October 27, 2004.

Clinic of Rheumatology, Medical University, Plovdiv, Clinic of Rheumatology, Medical University, Sofia, and Department of Maxillofacial Surgery, Medical University, Plovdiv, Bulgaria

Address for correspondence: M. Panchovska, MD, Clinic of Rheumatology, Medical University, Plovdiv, Bulgaria.