

Lymphoma in Sjögren's syndrome

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Introduction. Patients with Sjögren's Syndrome (SS) have an increased risk for developing lymphomas, in particular B cell non Hodgkin lymphomas (B-NHL). Previous reports have suggested that 1) malignant lymphomas occur in less than 10% of patients with SS, 2) they are usually low grade 3) lymphomatous change occurs more frequently in primary SS, and 4) patients with persistent glandular swelling, lymphadenopathy and extraglandular disease are at particular risk (1). This paper reports on some of the clinical, laboratory and histopathological features of 9 SS patients that developed lymphoma.

Methods. At the University of Connecticut Health Center we have a database of 255 SS patients. The majority of these are primary SS patients fulfilling modified European Criteria for SS (2), (modified by the use of; 1. Saxon test. 2. Gallium and technetium scans as tests for glandular inflammation and function). Nearly all of these patients have had a salivary gland lip biopsy. Patients are followed regularly and screened at least once a year for evidence of lymphadenopathy, hepato- and/or splenomegaly.

Results. Nine female SS patients have developed lymphoma, 8 with definite and one with probable SS (Table 1). Patients 1 and 2 have SLE and patient 3 has CREST. Four of 6 patients with primary SS had longstanding disease, a monoclonal spike and chronic swelling of parotid glands.

Pt.	SS	Antibodies	Site of lymphoma	Type of Lymphoma	Outcome
1. FA	2°	Ro, La, Sm, ANA, Rh factor	Pulmonary	P.D.L chemotherapy	Died 2 yrs after lymphoma
2. LV	2°	La, Sm, ANA Rh Factor, aCL	Posterior cervical node	L.C.I.L chemotherapy 2 courses	Progressed to stage II stable
3. GS	2°	Rh Factor, ANA	Parotid gland	M.A.L.T surgery	Doing well 3 yrs
4. K ⁺	1°	*	Gastric/pleural effusion, salivary glands	M.A.L.T chemotherapy	Doing well 2 yrs
5. AF	1°	Ro, La, Rh Factor	Multiple nodes	P.D.L chemotherapy	Doing well
6. GA	1°	Rh Factor	Submandibular mass	L.C.I.L chemotherapy	Doing well
7. JC	1°	Ro, Rh Factor, ANA	Retroperitoneal nodes	M.L. chemotherapy	Doing well
8. DO	1°	*	Lacrimal glands	M.A.L.T chemotherapy	Doing well
9. SM	1°	Ro, Rh Factor, ANA	Pulmonary	Marginal lymphoma	Just diagnosed

P.D.L Poorly differentiated lymphoma, L.C.I.L Large cell immunoblastic lymphoma,
M.A.L.T Mucosal associated lymphoid tissue, M.L. Malignant lymphoma, not defined

* None detected post lymphoma development

Conclusions. 1) Non-Hodgkin B cell lymphomas develop in a small number of patients with SS (3.5%). 2) Patients with lymphomas and secondary SS may not do as well as patients with primary SS and lymphoma. 3) Patients with longstanding primary SS and chronic glandular swelling appear to be at risk for malignant transformation.

References

1. Sem Arth Rheum 1998; 28: 80-87.
2. Arthritis Rheum 1993; 36: 340-47.