**SUPPLEMENTARY MATERIAL**

**Table S1 (Supplementary table on-line)**. Clinical, laboratory and histological features of patients developing lymphoma.

|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
| Patient | SÅ | SH | IN | GE | CO | UE | EV |
| Age at SS diagnosis | 69 | 50 | 52 | 55 | 46 | 47 | 62 |
| Age at lymphoma onset | 72 | 57 | 65 | 62 | 56 | 49 | 73 |
| Age at demise | n.a. | 57 | n.a. | n.a. | n.a. | n.a. | 76 |
| Diagnostic delay§ | 21 | 18 | 25 | 5 | 1 | 4 | 28 |
| Follow-up time after lymphoma diagnosis | 12 years | 4 months | 9 years | 6 years | 4 years | 17 years | 3 years |
| Treatment given for lymphoma | Radiation followed by 2 courses of chlorambucil and later followed by rituximab | Treatment attempts unknown (death 1992) | CHOP-R + intrathecal chemotherapy followed by high dose Methotrexate and autologous bone marrow transplantation | Radiation | CHOP-R | Radiation for the initial MALT. CHOP-R for the mantle cell lymphoma, followed by bone marrow transplantation | CHOP followed by chlorambucil + etoposid |
| Gender | Male | Female | Female | Female | Female | Female | Female |
| Lymphoma  (ICD7/10) # | 2001/C859  Low grade | 2001/C851  high grade | 2001/C851  high grade | 2001/C827  low grade | 2001/C838  high grade | 2001/C838  low grade | 2001/C859  high grade |
| Lymphoma type (WHO classification) | Follicular  B-cell lymphoma | Anaplastic large T-cell lymphoma | Diffuse large  B-cell lymphoma | MALT.  Lymphoplasmocytic/  Immunocytoma | Diffuse large B-cell lymphoma | MALT developing into mantle cell lymphoma | Diffuse large B-cell lymphoma |
| Primary localisation | Salivary glands | LNs, liver spleen, BM | Lungs | Tear gland/orbita | Stomach, LNs | Salivary glands | Stomach, liver, lungs |
| Salivary gland swelling | -\* | -\* | +° | + (tear gland) | +\* | +\* | +\* |
| Palpable purpura/skin vasculitis | -\* | +\* | +° | - | +\* | +° | - |
| Peripheral neuropathy | -\* | +° | +**°** | - | - | - | - |
| Enlarged LN/spleen | -\* | +° | +° | + | + | +° | +° |
| Associated predisposing condition | Psoriasis\*  *H pylori °* | - | Celiac\*  *H pylori°*  Hashimoto\* | - | Skin cancer  (MM) several | - | Breast cancer 6 months before lymphoma/radiation therapy |
| RF | -\* | -\* | +\* | - | + | - | - |
| Ro/SSA and/or La/SSB | +° | +° | +° | - | + | +° | - |
| ANA | +\* | +° | +\* | + | + | +\* | +\* |
| Cryoglobulinemia | -\* | -\* | +\* | - | +\* | - | ND |
| Leuco-/Lymphopenia | -\* | +° | +\* | (+) | + | +° | +° |
| Anaemia | -\* | -° | +\* | - | - | +° | - |
| C3 g/l | 0.98\* | 0.83\* | 0.78\* | 0.92\* | 0.94\* | 0.64\* | ND |
| C4 g/l | 0.27\* | 0.16\* | 0.18\* | 0.22\* | 0.09\* | 0.18\*0.09° | ND |
| CD4penia§§ | +\* | +\* | +\* | - | +\* | ND | ND |
| IgG g/l | 11.9\* | 23.0\* | 34.1\* | 11.00 | 18.6\* | 8.19\* | 13.2° |
| IgA g/l | 2.20\* | 1.00\* | 0.13\* | 3.38 | 5.5\* | 2.88\* | 1.89° |
| IgM g/l | 1.76\* | 1.03\* | 1.33\* | 1.23 | 4.3\* | 1.15\* | 1.17° |
| Monoclonality | -\* | -\* | -\* | oligo | + (2g) | -\* | -° |
| Salivary gland biopsy focus score ≥1 | Unknown/+ at re-evaluation | +\*/+ at re-evaluation | +\*/+ at re-evaluation | +\*/ (+) at re-evaluation (few and small foci) | +\*/+ at re-evaluation | +\*/+ at re-evaluation | Unknown/+ at re-evaluation |
| GC positive | + | + | + | - | + | + | + |

n.a. not applicable, §delay from first SS symptom to diagnosis of SS, \*evaluated at diagnosing SS, °developing during the course of disease before lymphoma development but not present or not analysed at first visit, #according to cancer registry, ND = not done, BM = bone marrow, LN = lymph node, MM = malignant melanoma, §§CD4-penia is defined as number of CD4+ T-lymphocytes below 0.3, % of CD4+ T-lymphocytes below 30, or ratio of CD4+/CD8+ T-cells ≤ 0.8.