ABSTRACT # 067

MEDIASTINAL SCLEROSING LARGE B-CELL LYMPHOMA. AN OVERVIEW BASED ON THE RESULTS OF A PATHOLOGIC TRIAL SUPPORTED BY THE INTERNATIONAL EXTRANODAL LYMPHOMA STUDY GROUP (IELSG)

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Background
PMBL has been the object of numerous reports, which have provided conflicting results in terms of phenotype, molecular characteristics, and histogenetic interpretation. During an IELSG study, 137 PMBLs were collected for extensive pathologic, phenotypic and molecular evaluation.

Results
At light microscopy, they mostly consisted of large cells with a predominantly diffuse growth pattern, varying degrees of nuclear polymorphism and clear to basophilic cytoplasm. The lymphomatous growth evoked a fibrous reaction with frequent compartmentalization. On immunohistochemistry, tumor cells showed the following phenotype: CD45+, CD20+, CD79a+, PAX5/BSAP+, BOB.1+, OCT-2+, Bcl-2+, CD30+, HLA-DR+, MAL protein+/−, Bcl-6+/−, MUM1/IRF4+/−, CD10+/−, CD21−, CD15−, CD138−, CD68−, and CD3−. The search for immunoglobulins (Ig) as well as for the corresponding m-RNA provided negative results. At molecular analysis, carried out in 45 cases, some novel findings were observed, which expand the spectrum of our knowledge on PMBL. In fact, more than half of the tumors displayed BC-L-6 gene mutations, which usually occurred along with functioning somatic IgVH gene mutations and Bcl-6 and/or MUM1/IRF4 expression. Frequent methylation of the MGMT gene was also observed.

Conclusions
The present study supports the concept that PMBL is mostly derived from activated germinal center or post-germinal center cells. However, the tumor differs from other aggressive B-cell lymphomas in that it shows defective Ig production in spite of an intact transcription machinery and lack of IgVH gene crippling mutations.