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## ABSTRACT # 362

### INTRAVASCULAR LARGE B-CELL LYMPHOMA (IVLBCL): INTERNATIONAL CONSENSUS ON DIAGNOSTIC AND THERAPEUTIC ISSUES

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#### **Background**

IVLBCL is a rare form of diffuse large B-cell lymphoma with preferential intravascular growth of malignant lymphocytes, aggressive behavior, and usually fatal course. IVLBCL often affects elderly patients with poor PS, elevated LDH serum levels, anemia, and B-symptoms, with possible differences in clinical presentation between cases diagnosed in Western Countries and Japan. Many characteristics of IVLBCL are poorly known and may be broader than those currently described.

#### **Methods**

On behalf of the International Extranodal Lymphoma Study Group, clinicians and pathologists interested in IVLBCL, coming from Western and Eastern countries, joined to reach a consensus on the most urgent unresolved issues in IVLBCL: to this end, a representative group of IVLBCL cases coming from Western countries and Japan were collectively analyzed. IVLBCL features were proposed both under clinical and pathologic standpoints.

#### **Results**

The consensus indicated that: IVLBCL have additional morphological criteria including definition of affected vessels, cell size (somewhat smaller cell may occur), peculiar sites of involvement (kidney, spleen and liver), and immunophenotype (in particular CD5 expression); difference in clinical forms of IVLBCL is driven more by the presence of haemophagocytosis rather than geographical origin of patients; "cutaneous variant", PS, stage, and therapeutic modality are independent prognostic variables; therapy is improved by anthracycline-based chemotherapy plus rituximab, CNS prophylaxis and consolidation with high-dose chemotherapy supported by autologous stem cell transplant, mostly in high-risk and young patients.

#### **Conclusion**

IVLBCL is a peculiar type of diffuse large B-cell lymphoma under pathological, immunophenotypic, clinical and therapeutical standpoints. An international prospective protocol, aimed to study both therapeutical and biological features of this disease is currently being designed.

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