Sistema Socio Sanitario
Regione Lombardia
ASST Sette Laghi

PRIMARY CENTRAL NERVOUS SYSTEM LYMPHOMA

Epidemiology

Primary CNS lymphomas (PCNSLs) in immunocompetent patients represent 2%–3% of all brain tumours. Most of these cases are DLBCLs and 10%–20% develop intraocular lesions.

Signs and symptoms

PCNSLs cause symptoms in most patients. These symptoms include behavioral or other cognitive changes, headache, confusion, nausea, and vomiting, seizures, weakness, sensory changes, such as numbness, tingling, and pain, weakness in the arms and legs difficulty controlling bladder or bowel functions, spots in the vision.

Diagnosis and risk stratification

Testing for PCNSL includes obtaining images of the brain through radiological examinations like MRI of the brain and examining a small piece of tumor by stereotactic biopsy, a small surgery done under general anesthesia. To complete diagnosis and staging, a lumbar puncture that allows to analyze the cerebrospinal fluid, is performed. A PET-scan is used to study the extension of the tumor. In some cases, a bone marrow biopsy is required. Variables with prognostic impact are age, performance status, LDH levels, involvement of deep regions of the central nervous system, and level of proteins in the cerebrospinal fluid.

Treatment

The treatment of PCNSL is mainly systemic, that means that a combination of immuno- and chemotherapy is administered. Our treatment approach relies on national and international guidelines i.e ESMO (https://www.annalsofoncology.org), NCCN (DOI: https://doi.org/10.6004/jnccn.2004.0028). There are two phases of treatment: induction treatment based on chemotherapy combined with immunotherapy (Rituximab) and consolidation treatment. For this second phase, there are different approaches: additional chemotherapy, high dose chemotherapy followed by stem cells transplantation and radiation therapy. The choice relies on variables like age of the patient and tumor extension. Intrathecal chemotherapy (chemotherapy administered directly into the cerebrospinal fluid) may be used in selected cases. Patients with unsatisfactory responses to standard therapy may be evaluated for clinical trials.