Primary central nervous system lymphoma is an uncommon form of extranodal non-Hodgkin lymphoma where malignant cells form in the lymph tissue of the brain and/or spinal cord.

The most common site for primary central nervous system (CNS) lymphoma is the brain. There may be more than one tumour present, although 60-70% of cases have a single tumour. Primary CNS lymphoma may also start in the layers that form the outer covering of the brain (meninges) or in the eyes (ocular lymphoma).

How does Primary CNS lymphoma affect the body?
The main symptoms arise from increased pressure within the skull (raised intracranial pressure). A blockage within the fluid-filled spaces in the brain can occur causing cerebral spinal fluid (CSF) to build-up, which causes raised intracranial pressure. The tumour itself may also be the cause of increased pressure. Raised intracranial pressure can cause headaches, nausea and vomiting and disturbances to vision. Other symptoms may include seizures, confusion, changes in behaviour and in personality. Weakness or paralysis of one side of the body may also occur as a result of a tumour on one side of the brain.

Who does Primary CNS lymphoma commonly affect?
It is most common in people aged in their 50s and 60s, but can occur at any age.

Do we know what causes Primary CNS lymphoma?
Whilst the exact causes of primary CNS lymphoma are unknown, a weakened immune system such as in people who have HIV/AIDS, or have had an organ transplant are at increased risk of developing this form of lymphoma, however the incidence in people with a normal immune system is rising significantly.

How is Primary CNS lymphoma treated?
Treatment for primary CNS lymphoma includes chemotherapy and often radiation therapy. Steroid therapy (e.g. dexamethasone) is often given before chemotherapy, as this helps reduce the swelling around the tumour, and may even shrink the tumour, reducing the raised intracranial pressure. Chemotherapy is given into a vein, and often also into the fluid surrounding the brain (intrathecally). Methotrexate is the drug most commonly given this way, and is also given in high doses intravenously—either on its own or in combination with other chemotherapy drugs and/or steroids. Radiotherapy uses high-energy rays to destroy the cancerous cells and can be an effective treatment for primary CNS lymphoma in younger people. It is used in combination with chemotherapy and steroid treatment. Surgery is only used to provide diagnostic tissue, and is never curative alone. Anticonvulsants may be given if seizures occur as a result of a tumour. If treatment is insufficient or if the disease recurs, some people are given a stem cell transplant using either their own stem cells (autologous) or those from a donor (allogeneic). New treatments are being researched all the time and people may be invited by their doctor to take part in a clinical trial to compare a new treatment or strategy with the best available standard treatment.


For more information, freecall 1800 620 420
email info@leukaemia.org.au or visit www.leukaemia.org.au

Last updated: July 2012