CHAPTER 14

Brain Lymphomas - Tumors Originating in the Head

Hope is an invaluable asset to all of us in coping with illness, as well as with the frustration of not being able to identify a single best treatment option. And hope becomes even more vital when affirmation as to the most appropriate measures for managing your disease does not exist.

Neal Levitan, Executive Director, Brain Tumor Society
IN THIS CHAPTER

BRAIN LYMPHOMAS

GENERAL QUESTIONS
What is a lymphoma?
How common is a primary central nervous system lymphoma?
Does location make a difference?

DIAGNOSIS
What are symptoms of a lymphoma in the brain?
What is the difference between a “T” and a “B” lymphoma?
What tests are needed for the initial evaluation?

TREATMENTS
A team of doctors to treat my lymphoma
Options before deciding on surgery

NEUROSURGERY
Medications before and during surgery

RADIATION THERAPY
How the role of radiation therapy has changed

CHEMOTHERAPY AND OTHER DRUGS
Why chemotherapy is given before radiation therapy
Immunotherapy - A new and evolving role
Steroids - The how and why they are used
My chances for longer life or cure with a lymphoma

Key search words
- lymphomas
- staging
- immunophenotype
- radiation
- internet resources
- metastases
- T and B cells
- steroids
- Rituxan®
- antibody therapy
- MRI
- tumor markers
- chemotherapy
- therapy options
GENERAL QUESTIONS

WHAT IS A LYMPHOMA? IS THE TYPE IMPORTANT?

Lymphoma is a tumor that comes from immune cells. There are two types: the Hodgkin and non-Hodgkin lymphoma. The former is extremely rare in the head. Lymphomas usually grow in lymph glands, bones, or bone marrow anywhere in the body. Those starting in the brain are called primary central nervous system lymphomas (PCNSLs). The brain has no lymph nodes, so we do not know where they originate in the brain.

HOW COMMON ARE PRIMARY CENTRAL NERVOUS SYSTEM LYMPHOMAS?

Twenty-five years ago, a PCNSL was a rarity; today they are more common. The frequency of lymphomas has increased ten-fold from 0.5 percent to about 5 percent of all brain tumors. Why? The increase is due to three groups: a) patients with compromised immune systems who are now living longer (e.g., cancer and AIDS patients), b) patients with lupus, rheumatoid arthritis, and bone marrow transplantation who are receiving immune suppressive therapies; and c) patients who are having (more frequent) biopsies. The latter has led to diagnoses that are more accurate.

DOES THE LOCATION MAKE A DIFFERENCE?

Yes. Location can affect the diagnostic tests and therapy.

• Lymphomas originating in the brain are still unusual, so it’s imperative that a search begin in all lymph node areas (neck, groin, chest, abdomen), since lymphomas usually start there.

• It is more suspicious for typical PCNSLs to appear in the frontal lobes, while metastases from elsewhere can be in any location.

• Tumors in the spine will require chemotherapy into the spinal canal, chemotherapy into a reservoir in the brain, or radiation along the spine.

• The location alone will usually not affect obtaining a biopsy or the effectiveness of later therapy (see the story of Gerald below).
DIAGNOSIS

WHAT ARE SYMPTOMS OF A LYMPHOMA IN THE BRAIN?

Typical symptoms reflect the area affected by the tumor. Most lymphomas in the brain grow in the frontal and temporal lobes. Symptoms include headache, vomiting, forgetfulness, difficulty in finding words, confusion, double vision, wobbliness (ataxia), weakness of a leg or arm, and sometimes facial weakness.

WHAT ARE THE DIFFERENCES BETWEEN A “T” AND A “B” LYMPHOMA?

These non-Hodgkin types are named after types of normal immune lymphocytes from which they develop. In normal lymph nodes, the outer area produces T cells that seek out and destroy germs, while B cells, produced in the inner area, are programmed (by the T cells) to make antibody proteins that protect against future attacks. This is what happens, for example, with successful immunization to tetanus or polio. Normal and tumor cells from these areas bear similar “marker” proteins (called the immunophenotype) for T or B cells. The T and B lymphomas respond to different chemotherapy and radiation combinations.

WHAT TESTS ARE NEEDED FOR THE INITIAL EVALUATION OF A LYMPHOMA?

Most neurooncologists (brain tumor specialists) and other physicians will evaluate a patient with a suspected brain lymphoma in the following manner with:

1. Thorough medical history, general physical and neurological examination.
2. Brain and spine MRI with/without contrast to visualize the brain and tumor.
3. CT scan of the chest, abdominal MRI, or ultrasound including lymph node chains, liver, and spleen (to exclude primary lymphoma elsewhere).
4. Complete blood counts, sedimentation rate, liver and kidney function tests, serum and spinal fluid levels of lactic acid dehydrogenase (LDH).
5. Biopsy (almost always indicated).
6. Analysis of cells in spinal fluid, when a biopsy is dangerous (rare).
7. Evaluation of the tissue specimen or spinal fluid by a hematopathologist (a pathologist who specializes in diseases of the blood and lymph glands).
8. Immune marker analysis (immunophenotype) on the lymphoma tissue.