

What You Should Know About Primary Intraocular Lymphoma

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Primary intraocular lymphoma is a rare, but severe ocular condition that frequently eludes correct diagnosis for months. A lymphoma is a cancer involving lymphocytes, one of the many types of white blood cells of the body. White blood cells are involved in immunity – protecting the body from infection by outside organisms. Lymphocytes come in many forms including B lymphocytes that make antibodies, T lymphocytes that help in recognition of foreign molecules called antigens that are carried on the surface of viruses and bacteria, and macrophages that engulf and destroy invading organisms. Primary intraocular lymphoma usually involves B lymphocytes.

Two Types of Lymphoma That Involve the Eye and Their Differences

From the standpoint of the eye, one can categorize lymphomas into two classes. The type we are discussing – primary intraocular lymphoma – usually starts under the retina, the nervous tissue lining the back of the eye, and then spreads into the vitreous gel that fills the eye. Simultaneously it may start in the brain tissue and spill cells into the spinal fluid. Thus, diagnostic efforts usually involve getting a biopsy of vitreous, doing a spinal tap to look for lymphoma cells in the spinal fluid, and obtaining an MRI scan of the brain and spinal cord to look for infiltration of the central nervous system with lymphoma.

A second type of lymphoma – systemic non-Hodgkin’s lymphoma – starts outside the eye and less commonly secondarily involves the inside of the eye. When the eye is involved, it is more common in this case to see pink tissue growing under the conjunctiva, fleshy nodules in the eyelid or orbit, and rarely mounds of lymphoma in the choroid, the layer of blood vessels between the white scleral coat and the retina.

Figure 1. Anatomy of the Eye

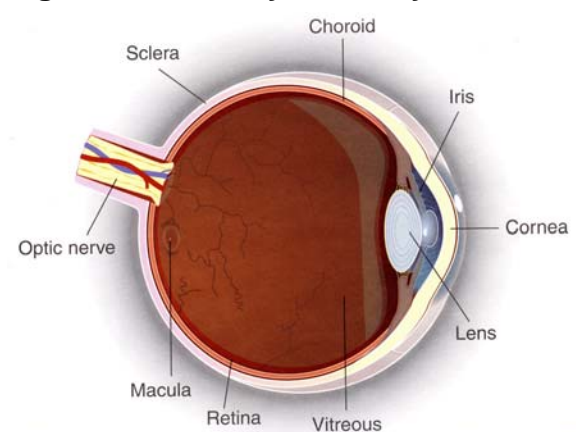
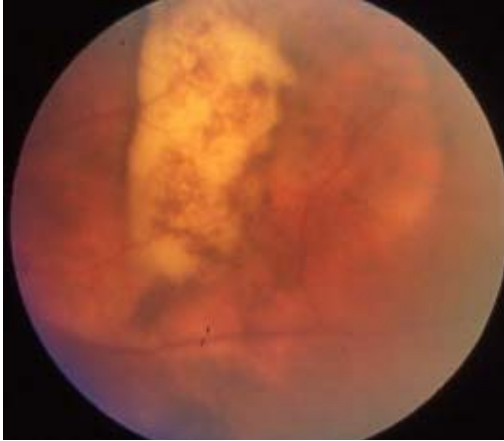


Figure 2. Normal Fundus



Figure 1 shows some of the ocular anatomy we have been discussing. Figure 2 shows the appearance of a normal back of the eye, as the ophthalmologist would see it. Figure 3 shows the appearance of primary intraocular lymphoma. A creamy mass is present under the retina.

Figure 3. Primary Intraocular Lymphoma



Both types of intraocular lymphoma are usually associated with lymphoma elsewhere in the body. Therefore, all patients with intraocular lymphoma need a work-up by a hematologist – oncologist to look for extraocular organ involvement.

Clinical Characteristics

Most patients with primary intraocular lymphoma are over 40 years old, with an average age of 60 years old. There is possible female prominence, but no race predilection. Usually both eyes are affected, although frequently at different times, thus both eyes are treated. In 80% of cases, the eyes are affected first, and the central nervous system later.

Treatment of Primary Intraocular Lymphoma

The ocular treatment consists of radiation therapy. Even when no evidence of central nervous system involvement is found, most patients receive systemic chemotherapy, and many receive injections of the drug methotrexate into the spinal fluid. Some patients receive radiation therapy to the brain and spinal fluid, especially when MRI scanning shows lymphomatous deposits in these organs. All patients require management by an oncologist.

Survival Statistics

Every case of primary intraocular lymphoma is different, and in some cases prolonged survival free of disease is possible. Taking all patients together, however, one is saddened to say that the prognosis is serious. On average, 30% of patients are alive five years after the diagnosis is made.

Final Comments

Primary intraocular lymphoma is a serious, often life threatening disease. With proper chemotherapy and radiation therapy, patients as a group are living longer. We can expect that further research may improve outcomes in the future.

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