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Eye of the Beholder

Many of my closest friends have known me for years, undoubtedly gazing into my eyes at some point in our acquaintance and not once noting the difference in size of my pupils. I find it fascinating when they have at last discovered what was “off” with my eyes and then ask for an explanation, until a few weeks ago I only partly had the answer. When I was four weeks old, my parents and a fleet of doctors discovered that I had Retinoblastoma; thus the legacy of Erin’s weird eyes begins. I hope that in the following paper the reader gains a better understanding of what Retinoblastoma is, how it is obtained, its signs, and recent courses of action in the study and future cure of this cancer.

As ophthalmological terms are not a part of common vocabulary, there might be many a term that perplexes the reader. For the sake of negating confusion, not wishing to befuddle the reader any more than he must, here are a list of terms that may arise in the following paper—terms found in Retinoblastoma International source:

Aqueous humor: Watery fluid which bathes and nourishes the front of the eye.

Bilateral retinoblastoma: Cancerous tumor(s) in the retina of both eyes.

Cornea: The clear outer part of the eye’s focusing system light entering the eye.

Iris: The colored part of the eye.

Metastasis: Spread of a cancer to other parts of the body.

Optic Nerve: The bundle of over one million nerve fibers that carries visual messages from the retina to the brain.

Prognosis: The overall outlook of treatment.

Pupil: The opening at the center of the iris that controls the amount of light.

Retina: The light-sensitive tissue lining the back of the eyeball.

Retinoblastoma: A cancerous tumor of the retina of the eye.

Unilateral retinoblastoma: Cancerous tumor(s) in the retina of one eye

One might wonder what exactly *is* Retinoblastoma? Retinoblastoma (reh-tin-oh-blast-oma) is a childhood cancer arising from immature retinal cells in one or both eyes. According to Retinoblastoma International the tumors develop in the human fetus, newborns, infants (premature and full-term), and preschoolers under age five. Ronald Aigotti likewise affirms that retinoblastoma is limited to children and in 1995 retinoblastoma occurred in 1 in 23, 000 live births, 6% of which had the tendency to be familial in nature (330).

Ronald Aigotti states that in many cases Retinoblastoma is discovered by age 2 and is rarely seen after age 6. Currently, the information stands that 87% of the children stricken with this disease worldwide die, mostly in under-developed countries. In developed countries, 97% of those who do live have moderate to severe visual impairment. In 1995 the statistics said that in the US approximately 300 children and adolescents younger than 20 years of age are diagnosed with retinoblastoma each year and that 5% of those retinoblastoma patients were associated with congenial defects such as mental retardation (330). Retinoblastoma now accounts for about 3% of the cancers in children under the age of 15 (Ret. International). Likewise according to Retinoblastoma International,

The tumors originate in the retina, the light sensitive layer of the eye, which enables the eye to see. When the tumors are present in one eye, it is referred to as unilateral retinoblastoma, and when it occurs in both eyes it is referred to as bilateral retinoblastoma. 60% of the cases involve only one eye (unilateral); the rest affect both eyes (bilateral). 90% of retinoblastoma patients have no family history of the disease and only 10% of newly diagnosed patients have other family members with retinoblastoma.

As to what causes the initial cancer, John Young states that the gene associated with retinoblastoma causes the tumor only when it is not working properly. The human body has two copies of this “tumor suppressor” gene that protect it against retinoblastoma and other tumors in the cells of their body. When both tumor suppressor genes stop or are prevented from working properly, retinoblastoma develops.

My father had retinoblastoma, and for the longest time, I did not know how he had inherited the disease, when my entire knowledge of the cancer revolved around the fact that I inherited it from him. So naturally, I assumed that it must be a completely inheritable disease, but this was a fallacy on my part.

Geoffrey Cooper says that retinoblastoma allows two forms to be distinguished: inherited and sporadic (non-inherited) retinoblastoma. Cooper likewise confirms my inquiries in stating that individuals with the inherited form of the disease transmit retinoblastoma susceptibility to half of their offspring, and while patients with inherited retinoblastoma frequently develop multiple tumors in both eyes, those with sporadic/non-

inherited retinoblastoma usually develop only one tumor in one eye. Children with sporadic retinoblastoma are also generally older than those with the inherited type when the tumor begins to develop (128).

Cooper maintains that because it is a fast spreading disease, early diagnosis is critical to successful treatment. Retinoblastoma can spread (metastasize) outside of the eye to the brain, the central nervous system—brain and spinal cord—and the bones. In such cases, chemotherapy is prescribed by a pediatric oncologist and is administered through the peripheral blood vessels or into the brain for months to years after the initial diagnosis (129).

As quick discovery of the disease is imperative to successful treatment, it follows that one should know the symptoms or signs that denote when a child may have retinoblastoma. As noted in Retinoblastoma International, Retinoblastomas are typically detected upon physical examination of the eye, in which the doctor can observe a whitened area of the pupil. In severe cases, one can view a white glow or glint in the pupil without the assistance of a microscope. The onlooker can also determine if a child has retinoblastoma if in a color photo the pupil is completely white or the eyes are crossed or misaligned.

Parents seem to have less control in this stage of treatment; they can only sit and watch their child's fate being handled in the many waltzing fingers and intellects of the doctors at hand. When asked what first went through her mind, my mother had this to say,

It's scary when your kid has any kind of disease, and then it's a problem because you want to love and protect them, and some things are just out of

your hands because you can't heal them yourself. One thing about retinoblastoma that is so scary is that you can't see it when it begins. I couldn't tell by looking at you that there was something wrong. Then I can't imagine what it would be like when saving your child's life calls for surgery, removal of the eyes. They were able to save your vision and your eyes (Erdos).

In regards to the question of treating retinoblastoma, Ronald Aigotti states that it is one of the few diseases in which biopsy is not required, but rather is discouraged as it can lead to fatal results. The physician can be virtually 100% positive of diagnosis with a few tests, such as CT scans and ultrasonic examinations of both eyes, even though the latter test is rarely implemented (331).

Aigotti further explains that cancer is staged in grades I – V based on the size and depth of the cancer within the eye, I having the best prognosis and V having the worst. To gauge which stage the retinoblastoma has entered a series of tests may be performed; it determines the extent of the cancer in the central nervous system and occasional distant spread into other organs. Staging Tests include CT scan of the orbit including the optic nerve, bone and base of the brain; x-rays of skull and bony orbits and lumbar puncture (spinal tap). When indicated or necessary, scans of entire brain, bone liver; complete blood counts, and chemistry profile can be helpful (330).

Prognosis is dependent on the grouping or stage of the cancer, the presence and appropriate treatment of said disease. In 1995, the mortality rate for retinoblastoma

hovered around 20%. There has been no substantial sustained change in retinoblastoma incidence after the 21-year period of 1975-1995.

To better explain the grouping Ronald Aigotti recorded the different stages of Retinoblastoma and how they relate to cure rates. Here is a list of statistics in relation to 1995: Group I- 80-85%; Group II- 65-70%; Group III- 60-65%; Group IV- 25-30 %; Group V- 15-20%. As one can see, the rates decrease as the severity of retinoblastoma increases, cases of retinoblastoma with distant spread to the brain or other organs have a very poor prognosis—chance of survival (331). Basically, if the cancer spreads near the brain through the optic nerve, chances go drastically down. However, Young states that survival for children with retinoblastoma (reflecting 1995 data) was quite favorable, with more than 93% alive at five years after diagnosis. Males and females had similar 5-year survival rates for the period 1976-94 (93-94%).

This elusive disease is still wanting a cure, yet treatments are available and are obviously successful due to the alive-ness of the author. Ronald Aigotti holds that the treatment of choice is the surgical removal of the entire effected eye along with 10-15 mm of the optic nerve (331). In my father's case of cancer, the doctors used this method of isolation in attempts to eliminate the cancer without loss of sight (Erdos). If both eyes are affected with retinoblastoma only the eye with the largest element of cancer is removed in order to attempt to preserve some sight in at least one eye.

According to Ronald Aigotti, most attempts to control cancer in the other eye are carried out with radiation therapy and chemotherapy. Some research facilities are trying to use radiation in the affected eye to preserve sight in that eye as well, but this approach is still under research investigation. Chemotherapy improves survival and cure rate

among patients (331). Though an outdated, by ten years, source, it holds true, as such was the case with my treatment. I did not undergo chemotherapy, but I did, however, undergo radiation therapy, in which a laser would eliminate the cancer from the pupil. Even after a treatment is finished, the parents, yet again, are robbed of their inherent duty, to protect their children from all harm at all costs. Again, all my mother could do when the initial treatment was over was wait and return year after year to affirm that the cancer had not returned. “There is always that aspect in the back of your mind that wonders if it will ever come back, which throws in the fear of it being unseen” (Erdos).

Another alternative to curing cancer, before the baby is even born, is abortion. As my mother so adamantly put it,

The doctors told us that there would be a 50/50 chance of you having cancer since your father had it in both eyes, eventually. They even brought me genetics counselors to talk with me about my “options” dealing with a cancer-prone child...meaning abortion. I have never been known to be angry, but that infuriated me. As if their answer to a problem was eliminating the child! (Erdos)

Honestly, I cannot remember much of the initial treatments waged throughout my first 6 months. I do remember returning every year until I was five, only to be strapped down, poked, prodded and coming home smelling of anesthesia. I remember the beds being large and the Ronald McDonald house in which my mother and I stayed many weeks at a time. Though I do not remember the treatments, I remember the yearly visits

with my doctor, who graciously saved my life. I am thankful to be in that small statistic of survivors.

Works Cited

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