What You Should Know About Idiopathic Juxtafoveal Telangiectasia
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Figure 1. Anatomy of the Human Eye

The retina is the neural lining of the back of the eye. Light is focused there and light energy is converted into a nerve signal, which travels to the brain via the optic nerve. The retina is only 200 microns thick, but it is richly supplied with a regular network of capillaries to bring oxygen and nutrients to the actively metabolizing cells. Figure 1 shows the anatomy of the eye. Figure 2 shows the regular capillary pattern of the retina. Idiopathic Juxtafoveal Telangiectasia (IJT) is a group of three related disorders in which the capillaries around the fovea, the center of the retina, are abnormally dilated and leaky. Idiopathic is a term used when the exact cause of the disease is unknown. Juxta means adjacent, thus juxtafoveal means adjacent to the fovea. Telangiectasia is a medical term that means dilated, thickened, and abnormally bent.

Three Categories of Idiopathic Juxtafoveal Telangiectasia

Group 1. These patients have easily visible abnormalities of the retinal vessels and show leakage with retinal swelling and fatty deposits in the retina. Most of the affected patients are male.
Group 2. This group is the most common. Either sex can be affected. Patients develop blurred vision in mid-life, usually of both eyes, together with a retinal sheen, presence of superficial retinal crystals, right-angle retinal venules, plaques of intraretinal pigment, and occasionally secondary abnormal blood vessels growing underneath the retina. The retina is thinner than normal. (1) The fluorescein angiogram, a test where food coloring is injected into the vein and multiple pictures are taken of the eye, will show staining of the paracentral retina. Figure 3 shows such a case.

Figure 3. Fluorescein angiogram of IJT

Group 3. This is the rarest group. These patients have abnormal gaps in the small blood supply around the fovea. They can have brain strokes, or other abnormalities of brain blood supply.

What Is The Cause of Idiopathic Juxtafoveal Telangiectasia?

The cause for any of the groups of this disease is still a mystery. It is suspected that a developmental abnormality is the cause, but no details are known. Occasionally twins or several members of a single family are affected, suggesting a possible genetic component in certain cases. However, this is the exception and not the rule.

What Can Be Done for Idiopathic Juxtafoveal Telangiectasia?

There is some suggestive evidence that diabetes mellitus is more prevalent in patients with IJT than in others, thus a test for diabetes is recommended for affected patients. Laser treatment has been tried, but generally found to be ineffective in improving vision. Case reports exist of mild improvement after photodynamic therapy using verteporfin, transpupillary thermotherapy, and injections of triamcinolone or bevacizumab into the vitreous cavity. (2) However, experience is preliminary and these treatments are not standard. Patients need to be aware of any new distortion in vision, which can signal secondary onset of a blood vessel growing under the retina. Looking at a grid of lines, such as an Amsler Grid, with one eye at a time, can help detect the occurrence of visual distortions. In this case, it is important to have an ophthalmologist check the eyes promptly, since laser treatment may be needed for this secondary consequence of IJT.
Final Comments

Idiopathic Juxtafoveal Telangiectasia does not blind, but it can cause distortion and blurry reading vision. Regular follow-up with an ophthalmologist experienced in this condition offers the patient the best chance of maintaining vision.

After you read this brochure, if you have further interest and questions, an excellent resource is the National Library of Medicine website called PubMed. It can be accessed via any search engine, or directly at this link, http://www.ncbi.nlm.nih.gov/entrez/query.fcgi. It includes an extensive database of reliable articles published in peer-reviewed medical journals from all over the world.

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Reference List
