Central Serous Chorioretinopathy

Central serous chorioretinopathy is a retinal disorder which affects the macula. It was first described in ophthalmology more than one hundred years ago. Essentially, it is an “idiopathic disorder” which means that the precise cause is unknown. Central serous is associated with an elevation (detachment) of the macula due to leakage of fluid from the circulation behind it (choroidal circulation). The leakage occurs through a defect in the tissue layer known as the retinal pigment epithelium. The retinal pigment epithelium is a single–celled layer that lies between the retina and the choroid (see anatomy page). This tissue layer normally serves to prevent fluid from the choroidal circulation from leaking under the retina. In central serous, fluid equilibrium is disturbed leading to leakage beneath the retina which elevates it to produce a macular detachment which distorts vision.

Although the precise pathophysiologic events leading to macular detachment are still poorly understood, the clinical manifestations begin with a disturbance in the retinal pigment epithelium which alters its normally impermeable state so that fluid leakage occurs, producing detachment of the retina itself. In this regard, the retina appears to be affected only secondarily. It is currently believed, according to research by our group, that the choroidal circulation develops a pool of extracellar fluid or edema. This intrachoroidal edema or leakage exerts pressure on the pigment epithelium, causing it to blister–up into focal or multifocal elevations known as serous detachments of the pigment epithelium. The blister disrupts or develops a mechanical opening, usually at the junction between its elevated and attached areas, permitting fluid to leak through the pigment epithelium beneath the neurosensory retina. This leakage is called “avascular” since it is not associated with the proliferation of an abnormal blood vessels (neovascularization) as is typical in the wet form of age–related macular degeneration. It is important to keep in mind that neovascularization may occur even in a patient with central serous chorioretinopathy as a secondary complication. So, while central serous chorioretinopathy is associated typically with non–vascular leakage, vascular leakage may evolve. One of the challenges in diagnosing central serous is distinguishing between these two types of leakage, especially in middle–age and older–age patients who are at risk for both central serous and age–related macular degeneration.

Clinical Features and Demographics

The syndrome is much more common in men who represent 85–90% of the cases. The onset is usually between the ages of 25 and 45, although, occasionally, a younger patient may be diagnosed with this disease. Older patients will continue to have signs of central serous when they developed it as a younger person, but some of the classic or typical features will be modified by age–related changes. Recently, Dr. Spaide in our group has identified a series of older patients with new onset of central serous. It appears that, in the past, these patients might have been diagnosed with the wet form of age–related macular degeneration.

Central serous patients in general tend to be slightly hyperopic (far–sighted) but not to have other eye disease. The condition is more common in Caucasians, Hispanics and Asians than in African–Americans who get this condition only rarely.

There has always been an impression that patients with central serous exhibit a characteristic personality. Patients have been observed to be energetic, dynamic, harried, pressured, and emotionally stressed. One of our earlier reports has identified the Type A Behavioral Pattern as a risk factor for the disease.

Clinical Findings

Patients usually present with a disturbance in central vision, either a gray zone or a blind spot in the central field or metamorphopsia which is waviness or undulation in the central portion of the vision. On clinical examination, the ophthalmologist may find one or more small blister–like elevations to the retinal pigment epithelium. In the acute stages, there is an overlying elevation of the neurosensory retina. Occasionally, patients who have detachments of the retina are asymptomatic, simply because the bubble of fluid does not involve the center of the macula or the foveal region. Signs of previous detachments of the retina can often be detected by the clinician. These are atrophic and pigmentary degenerative zones in the retina. When the neurosensory retina is elevated, it is displaced from its normal source of nutrition, the choroidal circulation. In time, the outer retina may become degenerated. Even when the macula resolves and is no longer detached, it does not function because of the degenerative change. (Thus, there is a rationale for reattaching the macula, if possible, see treatment, below.)

An important part of the diagnosis of central serous relies on the fluorescein angiogram. In the typical case, a leak can be demonstrated at the level of the retinal pigment epithelium as the fluorescein dye gradually passes into the subneurosensory retinal space. This is the hallmark angiographic feature of the disorder in its classic form. The leaks will vary, depending on the duration of the detachment and the nature of the fluid.

Some severe variants of the disease are seen in patients who have certain systemic diseases such as severe hypertension, collagen vascular disorders, blood dyscrasias, and organ transplant. The use of corticosteroids may also be a risk factor for central serous.

Treatment

There is no known medical treatment for the disorder. Most ophthalmologists urge patients to modify their behavioral patterns, to take a more relaxed or mellow approach to life. Our group has tried tranquilizers, antihistamines, non-steroid anti–inflammatory medications, and beta–blockers without success. One known method to reduce the duration of the detachment in an attempt to preserve the detached retina is laser photocoagulation. Under fluorescein guidance, the leak can be identified and treated with laser. Although some ophthalmologists believe laser treatment should be carried out early in the course of the disease, most feel that a conservative approach in

management should be entertained for a period of time before instituting treatment. For a leak that is remote to the center of the fovea, it is still reasonable to wait three to four months before considering laser treatment in the primary case. When the leak is close to the center of the macula (fovea), a more conservative approach to management is usually recommended. Laser treatment of such leaks has the risk of inadvertent damage to the center of the fovea or hemorrhage. These are rare complications, but there is a more certain possibility of a scotoma or a noticeable blind spot in the central visual field corresponding to the treatment site. When considering the potential risks vs. the potential benefits of laser treatment, the greatest rationale for treatment is the possibility of progressive loss of vision from detachment of the macula from prolonged detachment. Resolution of the detachment predictably occurs more expediently following treatment of the leak with laser. Each case must be approached individually. For example, in a patient who has had a previous detachment which has resulted in atrophic or degenerative scarring of the macula, earlier treatment of a leak in a subsequent detachment should be considered. The rationale is to reduce the duration of the second detachment to prevent further degenerative change in a patient who has already demonstrated loss from a previous macular detachment. Once degenerative changes have evolved, even resolution of the detachment in an accelerated fashion from laser treatment will not restore the vision completely. The longer the detachment, the greater the risk of permanent degeneration of the macula.

Prognosis

The prognosis for visual recovery in central serous is generally good. Usually, the pigment epithelial leaks closes spontaneously and the detachment resolves over a period of weeks to months. Most patients (greater than 90%) will retain vision of 20/30 or better in the affected eye. Although many patients may manifest subtle clinical findings in the opposite eye, most patients (greater than 80%) do not develop bilateral symptoms. Despite good acuity, some of these patients may still note some mild permanent abnormalities in the vision of the affected eye such as decreased contrast, mild distortion and decreased night vision. Reported recurrence rates are 20–30% although we have observed a higher rate among the patients referred to our group (perhaps a more severely affected group since we tend to be referred challenging cases). Weeks, months or even years later, new detachments may evolve. Each detachment runs the risk of further pigment epithelial and retinal damage. Some patients will experience progressive atrophy of the pigment epithelium and severe vision loss, which is permanent. Other patients may develop expanding detachments of the retina as fluid gravitates to detach the inferior retina.