

Eye Manifestations of Lupus And Sjogren's Syndrome

Based on a presentation by Dr. N. Kevin Wade at the BC Lupus Society Symposium held October 22, 2005.

Background

Dr. Wade completed his MD in 1998 at McGill University and his Internal Medicine Residency and subsequent Ophthalmology Residency at UBC in 1996. He obtained three years of further fellowship training in Uveitis and Neuro-ophthalmology in Montreal, San Francisco, and Vancouver. He has been practicing ophthalmology for the past 7 years and is on staff at Vancouver General Hospital. His current practice focus is cataract and general ophthalmology.

Overview of Eye Diseases in Lupus and Sjogren's Syndrome

Dr. Wade provided a broad overview of the anatomy of the eye, and some of the conditions that can be present as a result of dry eyes, lupus and Sjogren's syndrome, including:

- Inflammation of the arteries in the eye;
- Iritis, which occurs with inflammation of the inner linings of the eye;
- Cataract, which can occur as a result of inflammation in the eye, aging, and steroid therapy among others.
- Vitritis is where the jelly like substance behind the lens of the eye is inflamed;
- Diffuse uveitis refers to more than one level of the eye being inflamed when the eye is considered in terms of front, middle, and back;
- Periphlebitis, which involves inflammation above the veins in the eye;
- Scleritis, which is an inflammation of the white coat of the eye;
- Keratitis, which an inflammation of the cornea, can be very painful but is less likely in lupus
- Optic nerve inflammation. Most often the nerve may be involved by a stroke of the small blood vessels supplying the nerve – this is referred to as an ischemic optic neuropathy. Rarely optic neuritis is a primary inflammation of the nerve occurring in lupus or Sjogren's.

Dr Wade then elaborated on a few specific eye problems with lupus or Sjogren's syndrome:

Scleritis - Scleritis is the most frequent serious eye disorder found in lupus patients. Cataract is probably the most frequent cause of vision loss but not the most serious as it is easily remedied with surgery. Scleritis is generally visible when it involves the front of the eye. Scleritis is very painful and can cause permanent eye damage. It is possible that there will be ocular bruising around the eye in acute cases. When there is a severe inflammation of the eye, the tissue weakens as it heals. As a result, the eye is prone to future injury from the tissue loss. Rarely, the weakness in the wall of the eye can lead to loss of the eye.

Retinal vasculitis – Retinal vasculitis can cause serious damage to the retina, and is generally screened for regularly because of the possibility that it can develop in lupus. Retinal arteries can be infarcted (a clot causing a stroke of the retina) due to inflammation. Retinal damage will affect the field of vision and if it occurs in the center of the retina then central vision may be lost. This can lead to thrombosis in the blood vessels, which causes blockage and damage.

Choroiditis – (inflammation of the layer immediately behind the retina) can occur with loss of vision. Occasionally photographs of the back of the eye need to be taken with a dye injection in the arm (fluorescein angiography). This can show loss of blood flow in the choroidal layer which may be otherwise hard to see in the eye doctor's office.

Plaquenil toxicity - Drug treatment, in particular hydroxychloroquine or Plaquenil can cause pigmented deposits in the cornea and retina. This is more common with higher doses (more than 200 mg/day) and longer treatments times. There may be a ring of pigment which can cause loss of visual field. It is important to have yearly eye exams for toxicity if you are on Plaquenil. These eye exams should include colour vision testing, looking at a grid in the center of your vision, dilation of the pupil to see into the back of the eye, and central 10 degree visual fields. Glaucoma screening fields (usually 24 or 30 degrees in radius) are not sufficient to screen for Plaquenil damage.

Cataracts – Steroid therapy can cause cataracts. People usually notice glare which progresses to blurry vision. It may seem as if your glasses are not working properly. Night vision may also be reduced. The first sign of glare is difficulty driving into the sunset or difficulty with reflections from headlights on rainy nights.

Anterior Ischemic Optic Neuropathy – This may result from a stroke to the optic nerve it can occur in the front (anterior) or deeper (posterior) in the nerve as it leaves the back of the eye. Often the upper or lower half of vision is lost in that eye.

High blood pressure – High blood pressure can cause significant eye damage as a result of its short and long term effects on the optic nerve, retinal blood vessels, and choroidal blood vessels. Because lupus may affect the kidney it often results in high blood pressure problems. Occasionally, an eye examination may reveal the first evidence of high blood pressure because of retinal blood vessel and optic nerve changes.

Migraines – headaches and migraines are more common in people with lupus. This may be a result of blood supply problems in the brain – particularly in those who have antiphospholipid antibodies (proteins in the blood that may trigger clotting).

Dry Eye Syndrome

Dry eye syndrome is suspected from patient complaints of eye burning – often worse later in the day and diagnosed by measuring the tear production on a standardized paper filter strip inserted under the bottom lid of the eye, for five minutes. If tear production is 8 mm or less, the diagnosis of dry eye is confirmed. The most common cause of dry eye in lupus and Sjogrens is lacrimal gland inflammation. When the mouth is dry also it is called keratoconjunctivitis sicca.

Similar eye symptoms can be caused by meibomitis – a plugging of oil secretions in the eyelid glands. Many people have this condition without knowing it.

Meibomitis can cause red eyes, burning eyes in the morning and similar symptoms whereas true dry eyes are generally worse in the evenings. In essence, meibomitis occurs as a result of the loss of the normal oily coating in the tears, which prevents evaporation. As a result, tears will evaporate more quickly. Steroid tablets or eye drops may worsen meibomitis. Both dry eyes and meibomitis may be helped by warm wet compresses and artificial tear products such as Refresh Endura (one of the few artificial tear products to contain replacement oils and no preservatives).

Issues that may arise as a result of dry eyes include the following:

- Small erosions in the epithelium (corneal surface lining), which are very painful and may make it difficult to tolerate bright lights.
- A dry eye is more prone to infection.
- Inflammation of the lacrimal glands (tear gland) can cause dry eye as a result of accumulation of invading white blood cells. On a CT scan, one can often see that the gland is enlarged in Sjogren's syndrome. These white blood cells damage the gland and reduce tear production.

There are new theories with respect to the causes of dry eye. It is now thought dry eye is more of an inflammatory condition rather than merely a loss of tears. Altered secretions from the lacrimal gland inflame the ocular surface. This may cause pain which signals the brain and results in more inflammatory by-products developing in the tears. Basically, this small volume of highly concentrated tears can increase surface toxicity if not allowed to drain away. Because of this newer theory, ophthalmologists are generally using punctal plugs (thought to block tear outflow) less often. Some people may benefit from punctal plugs but many will not.

But, the question then arises -- if it is inflammatory why not use topical steroids? Steroids reduce local immunity so there is a rationale for their use. While oral steroids can be used it is rare to do so because of damage and loss to bone structure. Topical steroids are now occasionally being used to treat the inflammatory component of dry eye.

To diagnose Sjogren's syndrome, practitioners may look at the salivary glands in the mouth through a biopsy. If the salivary glands are inflamed with lymphocytes – it supports a diagnosis of Sjogren's syndrome. Unfortunately, lip biopsies can lead to a small area of permanent loss of sensation. For this reason, biopsies are not frequently done if the diagnosis is otherwise obvious.

Options for management of Dry Eye Syndrome

- Artificial tears – low or no preservative;
- Warm, wet compresses, 5 min and 3 times/day – can make eyes much more comfortable;
- Can take away medications which cause dry eye, such as antidepressants and antihistamines. An easy thing to do is to add a humidifier to your house;
- Topical fluorinated steroid, to be used carefully and eye pressure checks need to be done as some people develop glaucoma while on topical steroids;
- Increase water intake;

- Temporary punctal occlusion, but this is going out of favour with the medical community;
- Consider side shields on glasses;
- Moisture chamber glasses;
- Tarsorrhaphy (procedure whereby eyelids partially sewn together to decrease exposure of the eyeball to the air)
- Oral Salagen – this is an expensive drug but in low doses can be tolerated and will increase tear productions (it is also used for dry mouth).
- Avoid all preservatives in artificial tears if you use tears/drops more than 3-4 times per day – BAK can cause damage to corneal epithelial cells.
- One company has developed Restasis. This product treats the glands which produce tears. It is one of the few eye drop products which increases tear production. Unfortunately though, it is currently not available in Canada. We expect it to be released soon. In the US it costs about \$100 per month.
- Artificial tears are also starting to be developed which slow down evaporation. Another new product is Systane, which produces a dissolving barrier shield over the eye. This gel behaves as a temporary contact lens to slow down evaporation.

Cataract Surgery

Cataracts are more common in lupus patients. Cataract surgery is the most common surgery in Canada and is still the leading cause of blindness in the world. The most common indications are poor night vision, glare, or decreased vision affecting driving. Typically, a change in glasses prescription does not improve the condition. Other symptoms include colour loss and sometimes double vision in one eye. It is usually the vision requirements to continue driving which prompt many people to have an eye exam for diagnosis.

Cataract surgery is an 8 – 12 minute procedure, in which the cataractous lens is removed and an artificial lens replaces it within the capsule of the old lens. Surgery has a 99% chance of success, with a 1% combined estimate of complications. Surgery is covered by the Medical Services Plan for all residents of BC with an MSP number. The replacement lens lasts forever. A hard or rigid lens is paid for by MSP but most people choose a foldable lens which they pay for themselves. The foldable lens can be put in the eye through a much smaller incision (2.75 mm).

Glaucoma

Glaucoma is a disease of the optic nerve which affects the field of vision. Most people are unaware they have the disease when it is diagnosed. Ninety percent of glaucoma is of the open angle variety (this form does not prevent you from taking cold remedies warning you “do not take if you have glaucoma.” Ten percent is the “closed” angle variety. There is no absolutely normal eye pressure but higher pressures lead to faster optic nerve damage. There are a lot of new tools to study the optic nerve in glaucoma. HRT and OCT machines can be used to get an accurate measure of the optic nerve so that any damage can be picked up as soon as possible and eye drop treatment for glaucoma can be started as soon as possible. Corneal pachymetry (thickness of cornea) is often done to help explain abnormal eye pressure readings.

Reasons for IMMEDIATE Eye Evaluation

If you have following conditions, seek emergency eye evaluation:

- Severe pain in the eye, could be scleritis, keratitis, infection or angle closure glaucoma;
- “Flashes and Floaters”, usually means that the jelly-like substance filling the eye has detached from the retina and this can subsequently tear the retina or result in retina detachment which can cause permanent vision loss;
- Curtain or veil blocking eye, this can be an indication of retinal detachment or a stroke in the eye;
- Difference in size of eyes – proptosis is where an eye protrudes abnormally. If this is new it may result from thyroid eye disease or a growth behind the eye. Sometimes high or low eyelid positions may make an eye look more or less prominent; probably the worst cause of a new painful droopy eyelid is an aneurysm in the brain. This is a medical emergency.
- New strabismus (crossed eyes);
- New nystagmus (constant movement of eyes);
- Diabetes patients with decreased vision;
- Anyone with Type II diabetes should be evaluated within months of diagnosis.

Please speak to your regular ophthalmologist or optometrist if you have any further questions.

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