Case 21

History of Present Illness

The patient is a 42-year-old registered nurse, who is referred for eye pain and photophobia. She has a history of rheumatoid arthritis and mild hearing loss. At age 31 she began experiencing bilateral eye pain and headaches associated with severe light sensitivity. Despite having hearing loss, she said sound increased the pain in her forehead. She put blankets on the windows of her house and was only able to leave the house at night. She would experience a squeezing sensation in her eyes that moved to her forehead. She thought they were sinus headaches, and underwent sinus surgery even though the otolaryngologist expressed doubt that the sinuses were causative. She had no relief of the eye pain or improvement of her light sensitivity. Sometimes the pain was so bad that she would cry and vomit. While she felt that her eyes could be puffy, she denied any rhinorrhea, tearing, or redness of the eyes. She also had several stressors at that time including a new diagnosis of rheumatoid arthritis and marital discord. For several years she spent a lot of time in bed. She had to quit working and she wore sunglasses all day, inside and out. She had been treated intermittently in the past with TobraDex eye drops, but these no longer give her relief.

She saw a neurologist who gave her topiramate that caused mental slowing and another medication (that she did not know the name of) made her depressed. She was diagnosed with depression and treated with duloxetine, which helped her mood, but she discontinued this because she worried it caused sinus problems.

Her current symptoms are squeezing eye pain that radiates into her forehead with extreme light sensitivity, mild sound sensitivity, and nausea with rare vomiting. Sometimes she will have daily severe headache and photophobia. She is light sensitive every day all of the time in both eyes. Her eyes feel dry and scratchy every night and she also has dry mouth.

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Past medical and ocular history Rheumatoid arthritis Depression	*Past surgical history* Sinus surgery
	Family history Mother had "sinus headaches" Hypertension, depression
Medications Vitamin C and E and D and cod liver oil Oil of oregano Polymyxin B ophthalmic solution prn Tobramycin (tobrex) prn Dexamethasone 0.1% solution prn Valacyclovir prn	*Review of systems* Joint pain Dry mouth
Social history Married with three children 16 years of education; no smoking; no alcohol	

Examination

Acuity with correction Right eye: 20/20 Left eye: 20/20
Pupils Equal and there is no RAPD
Color vision (HRR) 6/6 OU
Stereo vision Excellent stereopsis: 8/9 circles on the Titmus test
Intraocular pressure Right eye: 15 mm Hg Left eye: 15 mm Hg
External exam Normal
Eye alignment Normal
Slit lamp examination She had Meibomian gland dysfunction in both eyes; normal anterior chamber and no cells were seen. She did have 2+ punctate epithelial erosion and tear film debris
Visual field Normal to confrontation
Fundus examination Normal optic disc, macula and retina
Neurologic examination Normal, except that she has allodynia on the forehead when it is touched. She also has mild hearing loss bilaterally.
Schirmer's test 4 mm OD and 5 mm OS; topical proparacaine improved her eye pain

Discussion

Neurologic Perspective: Dr. Digre

This patient presents with eye pain and photophobia, but she also has migraine headaches (see Case 19) and dry eyes (see Case 1). Both dry eyes and migraine can cause photophobia, and while she has been given eye drops in the past, she really had not understood how all of these symptoms could make each other worse. Most folks have a reason for photophobia and for some patients there may be more than one reason—such as dry eyes and migraine. My approach to diagnosing photophobia is this: first, a careful history is essential—I am looking for any central causes of photophobia such as meningitis, pituitary tumor or other clue. If so, I proceed with imaging (MRI) and possibly lumbar puncture. If not, I look for dry eyes. Dry eye symptoms are extremely common and we have found that patients with chronic migraine have more of these symptoms—even where there are no findings of erosions or decreased tear film (such as in our case). Treatment of dry eye symptoms may be helpful to reduce photophobia. I often will instill a drop of anesthetic to the eye, when eye pain is associated with photophobia to see how much the cornea is contributing to the symptoms. This helps with inflammation of the cornea, and corneal neuropathy in many cases. Then I think about the retina—is there hemeralopia (blindness from light) or night blindness, or trouble seeing the stars at night. A dilated examination can pick up retinitis pigmentosa or other retinal dystrophy associated with photophobia. I assess if there is excessive blinking to go along with blepharospasm (see Case 6)—another very common cause of photophobia and these individuals also may have dry eye symptoms. Finally, I am very careful to look for a headache history—since many patients have underlying migraine headache, which makes them more susceptible to photophobia. If I have not found a reason for photophobia, I start over, because there is usually a reason. Coming up with a diagnosis of the cause of photophobia is most important since treatment will frequently be directed toward those cause(s). In this case, I think she has photophobia due to migraine predisposition and dry eyes (Fig. 21.1).

I would recommend maximally treating the dry eyes. Sometimes with meibomian gland dysfunction, warm packs can be helpful. Frequent preservative-free tears can be helpful, and even ointment at night. While we discourage making the house darkened and wearing sunglasses indoors, FL41 tint which blocks blue light (which is the same wave length of the melanopsin pathway) and has been shown to exacerbate photophobia, seems to be helpful in both blepharospasm and migraine. When photophobia and eye pain are so severe, I have found gabapentin 100 mg three times daily and working up to a dose tolerated and efficacious is also helpful. Correcting the migraine component may also be important—treatment of migraine is discussed in Case 19.

The cause of photophobia is not completely understood, but it is very clear that one does not need vision. The melanopsin pathway of intrinsically photoactive ganglion cells synapse in the thalamus and connect with trigeminal afferents from the

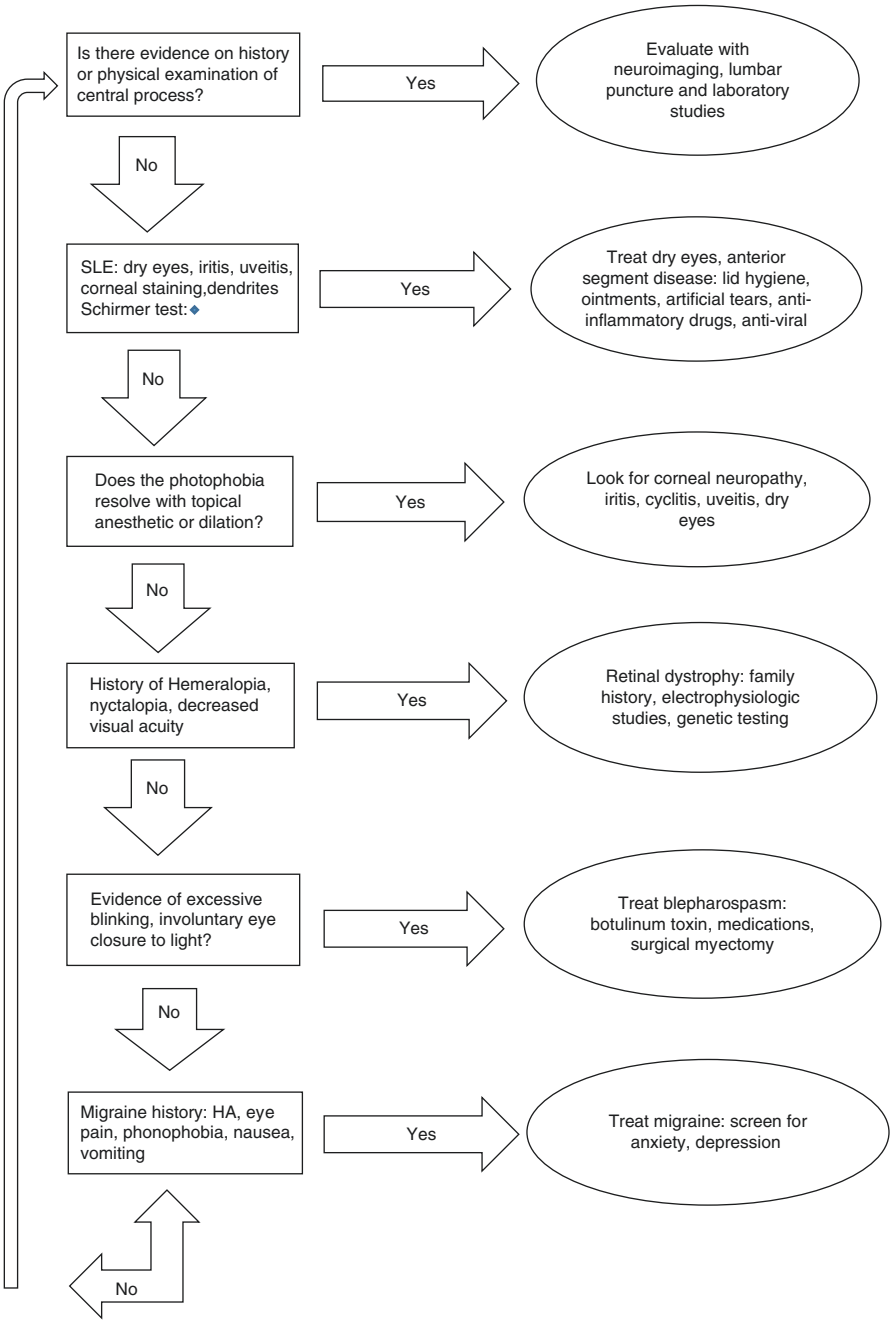

Fig. 21.1 An approach to the patient with photophobia (adapted from Digre and Brennan)

dura and presumably the eye causing the sensation of pain. Why some individuals have lower thresholds is unknown, but it is clear that individuals with blepharospasm and migraine have lower thresholds. One can understand why inflammation from iritis, conjunctivitis, scleritis, and so forth in the eye could affect trigeminal afferents directly and contribute to photophobia. We are beginning to understand why some individuals retreat into the darkness of their homes, and become very depressed too. Clinical studies have shown individuals with interictal photophobia in migraine had more depression and anxiety. Furthermore, newly born fetal animals with only the melanopsin pathway active show signs of anxiety and have increased expression of C-fos in the amygdala when exposed to light at an early stage. These studies show that there may be an emotional component to photophobia. It probably contributes to many providers diagnosing factitious disease among individuals with sunglasses in their waiting room.

Ophthalmic Perspective: Dr. Lee

Wearing sunglasses indoors is not socially acceptable because folks cannot see someone's eyes. Additionally, the sunglasses dark adapt the individual and make them more light sensitive (similar to when we non-photophobics walk outside of a movie theater on a sunny day). Tinted lenses are much more acceptable and the patient generally can see better indoors with them. Keep in mind that a patient can order a light or a heavy FL-41 tint. Many will order a light tint for situations with fluorescent lights and heavy tint for outdoors. There are also wraparound or cocoon frames that block the light from getting in from the sides. Finally, I like to tell patients to try smart lightbulbs. These LED lightbulbs connect to the internet and are controlled by smartphones. You can change the color and set the brightness to something that is more comfortable. There are also some optometrists and opticians who can dispense tinted contact lenses. However, contacts are more difficult if dry eye is a significant component of the photophobia.

Anything that irritates or inflames the cornea or uvea can cause photophobia, which will generally be reflected in a red eye. However, sometimes the redness is subtle or absent. Patients can have uveitis (see Case 12). This will generally cause some blurring of the vision. Definitively evaluating for uveitis requires a slit lamp and dilated fundus examination. I do not know who gave her Tobradex eyedrops, but hopefully it was an eye doctor. The absence of redness or tearing argues against infection so I'm not certain why give her the antibiotic portion of the drops. As we all know, steroids treats a lot of things including dryness and inflammation but it can lead to cataract, high intraocular pressure, or worsen corneal infection. Anyone but an eye doctor should probably avoid prescribing steroid eye drops for more than a week.

Finally, this woman has allodynia of her forehead. This suggest some type of neuropathic pain. Hence, a trial of an anti-epileptic such as gabapentin may be reasonable.

Non-ophthalmic/Non-neurologic Perspective

The primary care physician will definitely need help from an ophthalmologist since making the diagnosis of dry eyes can be challenging. Sometimes the eyes will not even appear red or injected. The primary care physician will feel more comfortable treating the migraine component. For all patients with photophobia, it is reasonable to ask the patient to visit their optical shop and try to purchase FL-41 tinted lenses**.**

Follow Up

We did check Sjögren's antibodies, which were negative. We recommended FL-41 tinted lenses and this improved her symptoms markedly. This patient began using tears during the day and ointment at night. She was instructed that migraine can be worsened by her dry eyes and that some of her dry eye symptoms can accompany chronic migraine. She improved—she is still light sensitive but is able to control symptoms by controlling the dry eyes and treating her migraine and using the tinted lenses. We started her on gabapentin 100 mg at bed time and increased her dose weekly to a tolerated maximum dose of 300 mg three times daily. We suggested that she work with her ophthalmologist on the dry eyes and that she may even need punctual plugs. *Final diagnosis: photophobia.*

For Further Study

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