Case 10

History of Present Illness

A uveitis specialist sent a 13-year-old boy for eye pain. He had normal development. He started with eye pain at age 4 and was treated for allergies for 2 years. At age 6, he was diagnosed with non-granulomatous uveitis and was evaluated for systemic disease but none was found. He started having eye pain—usually one eye at a time occurring 1–2 times each year from age 6 to 12. He was treated with naproxen and prednisolone acetate drops for minor ocular inflammation. In between his bouts of inflammation, he was pain-free. At age 12 he developed episcleritis in the right eye and intermediate uveitis. Treatment with meloxicam and systemic prednisone completely quieted his eyes down. He had an orbital ultrasound and MR scan looking for orbital inflammation, but none was found. His work up included normal CBC, CMP, ESR, lysozyme, ACE, ANA, quantiferon gold, HLA B27, Serum IgG4, ANCA, and MPO. He had a negative RPR, FTA, toxoplasmosis, and toxocara titer. He was seen by a rheumatologist who started methotrexate and began tapering the steroids. When steroids were tapered to 5 mg, the eye pain recurred without increased ocular inflammation. Repeat ultrasound showed minimal scleral thickening. An orbital specialist evaluated him and no orbital disease was found. He was referred to neuro-ophthalmology for further evaluation since he had eye pain and no active uveitis or episcleritis.

The pain was a steady, constant ache in the left eye—he had no headache. He had no light or sound sensitivity and it did not worsen with activity, but sometimes the eye would tear. The pain lasts hours to days; and a cool rag over the eyes sometimes worsens it. A burst of 60 mg of prednisone would quiet the pain, but when he tapered below 15 mg, the pain would recur. Most of the time, it would occur in the left eye but he had bouts of right eye pain. His vision he says is normal.

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Past medical and ocular history Non-granulomatous uveitis	*Past surgical history* Inguinal hernia repair
Medications Calcium/Vitamin D Cellcept 500 mg Prednisone 15 mg	*Family history* Mother with migraine
Social history 7th grade	*Review of systems* Otherwise negative

Examination
Acuity with correction Right eye: 20/20-2 Left eye: 20/20
Pupils 5 mm OD, 6 mm OS in darkness; 4 mm OD, 5 mm OS in light; No RAPD
Color vision (HRR) 8/10 OD and 9/10 OS
Stereo vision Normal—9 circles by Titmus Test
Intraocular pressure Right eye: 17 mmHg Left eye: 17 mmHg
External exam Normal
Eye alignment Normal
Slit lamp examination One deep scleral vessel mildly enlarged left eye, otherwise normal without cell or flare
Visual field Normal to confrontation
Fundus examination Normal optic disc with 0.2 cup to disc ratio; normal macula and vessels
Neurologic examination Normal neurological examination

Discussion

Neurologic Perspective: Dr. Digre

My first thought was—is this migraine coming on when the patient has a history of inflammatory uveitis. His mother had migraine and he could not read in the car (often seen in migraine). However, he had NO headache whatsoever, no migraine features of light and sound sensitivity, no nausea, no worsening with activity. I also thought about a form of cluster or autonomic cephalgia, but the pain could be on the right or left side, lasted hours to days and completely went away after being on high dose steroids.

We also wondered about scleritis or episcleritis, but his imaging (MR, ultrasound) were said to be normal. He had no active uveitis. His intraocular pressures were normal. I tried him on indomethacin in case this was an unusual case of hemicranias continua. There was only a partial response.

Ophthalmic Perspective: Dr. Lee

Recurrent uveitis would be the most common situation causing eye pain, but the slit lamp examination was normal. However, I think inflammation of "something" seems likely given the steroid responsive nature of his pain.

Episcleritis and anterior scleritis cause eye redness, but episcleritis is not very painful while scleritis often becomes increasingly painful. If it is not clear, then a drop of 10% neosynephrine will blanch the vessels in episcleritis but will not affect deeper vessels in scleritis. Why is this important? As a general rule, episcleritis is not associated with a systemic disease. Scleritis can be idiopathic or associated with a systemic condition and also lead to visual loss.

In the absence of eye redness, then one could consider posterior scleritis or even orbital inflammation. Often inflammation of the orbit will cause orbital signs such as diplopia and ophthalmoplegia, proptosis, chemosis, and eyelid edema. Sometimes, myositis will cause eye pain with eye movements with a white and quiet eye.

Non-ophthalmic/Non-neurologic Perspective

Scleritis is not a frequent cause of eye pain. This is a situation where having an ophthalmologist examine the patient can be very helpful. Think about scleritis or eye inflammation in patients with autoimmune disease who have new onset of eye pain or red eye.

Follow Up

This is when I went back to the MR scan to review carefully**.** The MR scan showed faint enhancement of the sclera and also the perioptic nerve sheath (Fig. 10.1). The indomethacin helped a little, but did not stop the eye pain. We also repeated his ultrasound, and now he had classic findings of posterior scleritis. Posterior scleritis will only be visible on imaging of the posterior globe—as in this case on the MR scan.

Table 10.1 gives the ICHD 3 beta criteria for scleritis and eye pain/headache. Scleritis can often be misinterpreted as one of the autonomic cephalgias. But as in this case, indomethacin was only partially helpful. Scleritis can occur in children as well.

In a large review of scleritis, Lavric et al. reported that scleritis was more common in women (71%), less likely to be bilateral (15.8%), and often associated with autoimmune disorders such as rheumatoid arthritis, systemic lupus erythematous,

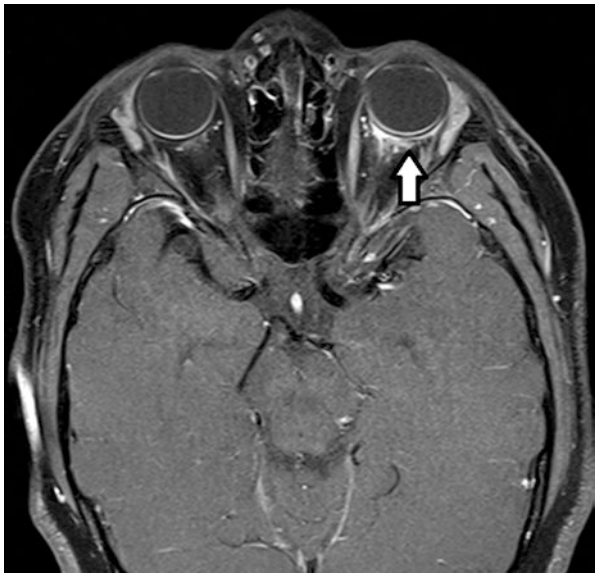

Fig. 10.1 Axial, postgadolinium T1 MRI of the orbits shows mild enhancement of the posterior sclera in the left eye (*arrow*). There is also perineural enhancement at the nerve/sclera junction

Table 10.1 ICHD 3 beta headache attributed to ocular inflammatory disorder

Diagnostic criteria:
A. Periorbital headache and eye pain fulfilling criterion C
B. Clinical, laboratory, and/or imaging evidence of ocular inflammatory diseases such as iritis, uveitis, cyclitis, scleritis, choroiditis, conjunctivitis, or corneal inflammation
C. Evidence of causation demonstrated by at least two of the following:
1. Headache has developed in temporal relation to the onset of the ocular disorder
2. Either or both of the following:
(a) headache has significantly worsened in parallel with worsening of the ocular disorder
(b) headache has significantly improved or resolved in parallel with improvement in or resolution of the ocular disorder
3. Either or both of the following:
(a) headache significantly improves with topical application of local anesthetic agent to the eye
(b) headache is aggravated by pressure applied to the eye
4. In the case of a unilateral eye disorder, headache is localized ipsilateral to it
D. Not better accounted for by another ICHD-3 diagnosis
According to the ICHD 3Beta—Description: Headache caused by ocular inflammatory conditions such as iritis, uveitis, scleritis or conjunctivitis and associated with other symptoms and clinical signs of the disorder
Headache Classification Committee of the International Headache Society. The International Classification of Headache Disorders: 3rd edition (beta version). *Cephalalgia*. 2013;33:629–808

angiitis and or infection. Relapses occur in about 1/3. Pain in the eye is present in over half; fewer (about 1/3) have blurred vision and light sensitivity is not common. Sometimes one can see subtle disc swelling and even folds in the retina and serous retinal detachment in posterior scleritis. Occasionally there can be longer lasting pain (neuralgiform type) that challenges treatment.

In this case, he could not be tapered below 15 mg prednisone daily. If a patient cannot get below 7.5 mg prednisone daily, then we would consider asking a rheumatologist to institute a steroid-sparing agent. The patient might also benefit from a periocular steroid injection to reduce the systemic load. *Final diagnosis: posterior scleritis*.

For Further Study

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