Identifying the Culprit: A Case Study of Retinal Vasculitis and HLA- B27

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Abstract:

A 16 year old female presents for a routine exam with no visual complaints. Dilation reveals vascular sheathing in the peripheral retina. She’s referred to a retina specialist with a suspected diagnosis of retinal vasculitis.

1. Case History
   a. Patient Demographics: 16 year old Hispanic female.
   b. Chief Complaint: No visual complaints. She is interested in trying contact lenses this year, due to glasses becoming bothersome during sports.
   c. Ocular History: Compound Myopic Astigmatism, bilateral.
   d. Medical History: None
   e. Medications: None
   f. Other: None

2. Pertinent Findings
   a. Clinical: Vascular sheathing in the peripheral retina of the left eye, no vitritis or uveitis present.
   b. Physical: The patient has not had any health issues recently.
   c. Laboratory Studies: Angiotensin Converting Enzyme, Antinuclear Antibody, Complete Blood Count with Differential, C-Reactive Protein, Erythrocyte Sed Rate, HLA B27, and Lyme Serology.
      i. All laboratory studies were normal, except it was found she is HLA B27 positive.
   d. Radiology Studies: Macula OCT showed an intact foveal contour in both eyes.
   e. Other: Fluorescein Angiogram revealed no evidence of leakage or staining in the area of the retinal sheathing.

3. Differential Diagnosis
   a. Primary/ Leading: Idiopathic Retinal Vasculitis, possibly due to previous inflammation from HLA- B27.
   b. Others:
      i. Retinal Vasculitis secondary to Tuberculosis.
         1. This differential diagnosis is higher on the list due to the patient traveling outside the country on an annual basis.
      ii. Most common causes of retinal vasculitis:
         1. Syphilis
         2. Sarcoidosis
         3. Pars Planitis
         4. Sickle Cell Disease
iii. Less common causes of retinal vasculitis:
   1. Multiple Sclerosis
   2. Eales Disease
   3. Viral Retinitis (eg, HIV, herpes)
   4. Behcet Disease
   5. Fungal Retinitis
   6. Bacteremia

4. Diagnosis and Discussion
   a. Elaborate On The Condition:
      i. Retinal vasculitis is characterized by ocular inflammation which involves the retinal vasculature.\(^{(2)}\)
      ii. Retinal vasculitis is rarely reported in association with HLA-B27. Retinal vasculitis associated with HLA-B27 showed primarily mild peripheral vasculitis.\(^{(1)}\)
      iii. Complications of retinal vasculitis associated with HLA-B27, includes macular edema and serous retinal detachment.\(^{(5)}\)
      iv. In one study it was found that retinal vasculitis due to HLA-B27 did not occur as an isolated phenomenon, but instead always developed with concurrent anterior or panuveitis.\(^{(7)}\)
      v. It is still unknown if retinal vasculitis is a rare manifestation of HLA-B27 or a coincidental finding. A larger prospective study would need to be done to further analyze this.
      vi. In one small cohort, study showed that the finding of retinal vasculitis with panuveitis may occur in the wake of HLA-B27 associated disorders and may in fact represent an infrequent manifestation.\(^{(7)}\)
      vii. The cause of poor vision associated with retinal vasculitis is multifactorial, but a significant contributory factor is cystoid macular edema.\(^{(1)}\)
      viii. When vision loss continues even with appropriate treatment, this is likely due to retinal ischemia. Retinal vasculitis with retinal ischemia has a significantly worse visual outcome than those with non-ischemic retinal vasculitis.\(^{(1)}\)
      ix. Inflammation induced retinal vasculitis can lead to a proliferative vascular retinopathy, which can produce vitreous hemorrhage, traction retinal detachment, rubeosis iridis, and neovascular glaucoma.\(^{(1)}\)
      x. Patients that present with idiopathic retinal vasculitis, are typically young adults with no signs or symptoms that suggest an underlying system or ocular disease.\(^{(2)}\)
   b. Expound On Unique Features: Retinal vasculitis is rarely reported in association with HLA-B27. In addition, the young age of the patient who presented with retinal vasculitis makes it a unique case.

5. Treatment, Management
   a. Treatment and Response to Treatment: No treatment necessary at this time, due to no leakage at the site of the peripheral retinal vasculitis. The patient will return for a follow
up examination with myself in around 6 weeks, and a repeat dilated exam in 3-4 months with the retinal specialist.

b. Refer to Research When Appropriate:
   i. The goal of treating retinal vasculitis is to quiet the intraocular inflammation in order to prevent vision loss and long term complications.3
   ii. When there is mild disease and visual acuity is 20/40 or better then therapy may not be required.3
   iii. Initial therapy consists of corticosteroids. Periocular corticosteroids are useful in patients with unilateral disease or moderately severe inflammation. Oral corticosteroids are used for patients with moderate to severe retinal vasculitis that is bilateral, and a marked reduction in visual acuity.3
   iv. Some patients may not response to the corticosteroid therapy, or may have adverse reactions and are unable to continue with them. In these cases, steroid sparing immunosuppressive therapy may be helpful.3
   v. Limited studies have been done using steroid sparing immunosuppressive therapy. Cyclosporine and azathioprine are the most common used for the treatment of retinal vasculitis.6
   vi. Laser photocoagulation and vitreoretinal surgical procedures may be necessary to treat complications associated with retinal vasculitis.3

6. Conclusion
   a. Clinical Pearls:
      i. Retinal vasculitis found on an exam warrants additional testing. The importance of distinguishing between retinal vasculitis due to an infectious versus non-infectious etiology helps to determine which treatment would be most beneficial.1
      ii. Laboratory workup should be ordered based on the differential diagnosis, which is derived from a thorough case history, review of systems, and physical examination.3
      iii. The absence of any diagnostic clues from the history and physical examination makes idiopathic retinal vasculitis the most likely. The initial tests recommended include erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), complete blood count (CBC) with differential, fluorescent treponemal antibody absorption test (FTA-ABS), Lyme and toxoplasmosis titers, chest x-ray (CXR), angiotensin converting enzyme (ACE), antinuclear antibody (ANA), rheumatoid factor (RF), basic chemistry panel, urinalysis, human leukocyte antigen (HLA) typing, human immunodeficiency virus (HIV) testing, PPD, and fluorescein angiogram.4
      iv. A fluorescein angiogram should be ordered in cases of retinal vasculitis. It is a sensitive technique and will frequently show more extensive vasculitis than what is seen during examination.1

7. Bibliography


