Title: Worm in the Eye: Diffuse Unilateral Subacute Neuroretinitis Likely Caused by *Baylisascaris procyonis*

Abstract:

Diffuse unilateral subacute neuroretinitis (DUSN) is a rare condition that usually results in severe vision loss. We report a case of DUSN presumably caused by the raccoon roundworm, *Baylisascaris procyonis*, in New York City.

I. Case History

1. **Patient demographics**
   a. 48 year old Hispanic male

2. **Chief complaint**
   a. Unilateral acute painless vision loss in the right eye of 3 days duration.

3. **Ocular / medical history**
   a. Pertinent Ocular History:
      i. Unremarkable
   b. Pertinent Medical History:
      i. Hypertension controlled with medications

4. **Medications**
   a. Unknown anti-hypertensive medications

5. **Other salient information**
   a. Patient works at a demolition site in New York where raccoons can be found in the area. He reports that dust got into his eyes as he was moving woods and demolishing a house filled with raccoon droppings prior to the onset of the acute vision loss in the right eye.

II. Pertinent findings

1. **Clinical**
   a. Entering visual acuity uncorrected:
      i. Right eye: 20/400 No improvement on pinhole
      ii. Left eye: 20/20
   b. Pupils: PERRL (+) APD right eye
   c. Extra ocular muscle motilities: Full without restrictions both eyes.

2. **Physical**
   a. Anterior Segment Exam:
      i. Unremarkable without evidence of cells in the anterior chamber both eyes
   b. Goldmann Tonometry:
      i. Right eye: 11mmHg
      ii. Left eye: 13mmHg
   c. Dilated Posterior Segment Exam:
      i. Vitreous: Asteroid hyalosis without evidence of vitritis both eyes
      ii. Optic nerve:
         1. Right eye: 0.35 pink and distinct
2. Left eye: 0.35 pink and distinct
   iii. Posterior pole / macula:
      1. Right eye: diffuse RPE mottling and choroidal changes throughout posterior pole and macular area
      2. Left eye: 2.5 DD chorioretinal atrophy at end of inferior temporal arcade
   iv. Normal vasculature both eyes
   v. Periphery:
      1. Right eye: RPE mottling with pigment clumping, no holes or breaks 360
      2. Left eye: no holes or breaks 360

3. Laboratory studies
   a. CBC with differentials
      i. Negative for eosinophilia
   b. Low C reactive protein and low sedimentation rate
   c. Laboratory tests were negative for:
      i. Toxoplasma IgG and IgM antibodies
      ii. Lyme antibodies
      iii. QuantiFERON-TB Gold
      iv. RPR and FTA-ABS
      v. HLA-A, B, C typing
   d. Patient returned to our clinic after being lost to follow-up for one month. At the 1-month visit, he presented with severe optic nerve pallor, attenuation of arterioles and diffuse RPE degeneration in his right eye with light perception vision. After ruling out infectious, inflammatory and compressive etiologies, anti-recoverin and anti-alpha-enolase antibodies were ordered which returned negative.

4. Radiology studies
   a. At the 1-week retinal consult, mild optic nerve head pallor was noted on fundus examination. MRI of brain and orbits with and without contrast were ordered to rule out compressive etiologies. Imaging results came back unremarkable.

5. Others
   a. Optical coherence tomography of the macula:
      i. Right eye: compromise of the IS/OS junction with diffuse retinal pigment epithelium stippling
      ii. Left eye: all retinal layers intact
   b. Fluorescein angiography at the 1-week retinal consult shows evidence of diffuse loss of retinal pigment epithelium in the right eye, which manifested angiographically as window defects.
   c. Careful review of color fundus photographs from previous visits revealed a live nematode and pigmented tracks within the posterior pole of the patient’s right eye.

III. Differential diagnosis

1. Primary/leading
   a. Diffuse unilateral subacute neuroretinitis
2. Others
   a. White dot syndromes
      i. Multiple evanescent white dot syndrome (MEWDS)
      ii. Acute posterior multifocal placoid pigment epitheliopathy (APMPPE)
      iii. Multifocal choroiditis
   b. Ocular toxoplasmosis
   c. Presumed ocular histoplasmosis
   d. Sarcoidosis
   e. The late stages of DUSN may be confused with retinitis pigmentosa, toxic retinopathy, occlusive vascular disease, and melanoma-associated/cancer-associated (MAR/CAR) retinopathies.

IV. Diagnosis and discussion

1. Elaborate on the condition
   a. Diffuse unilateral subacute neuroretinitis is an ocular parasitic infection that usually affects children or young healthy adults.
   b. It is believed that the disease is caused by nematodes of at least two different sizes. The smaller nematodes, measuring 400 to 1000um in length, are documented mainly in the southeastern United States and have been suggested to be the dog hookworm, *Ancylostoma caninum*. The larger nematodes, measuring 1500 to 2000um in length, have been reported in the north Midwestern United States and are believed to be the raccoon roundworm, *Baylisascaris procyonis*.
   c. DUSN typically presents as an insidious onset of unilateral paracentral or central scotoma. However, bilateral cases have been described in literature although uncommon.
   d. The identification of a worm is the gold standard in making the diagnosis of DUSN. Serologic studies for parasites, stool examinations, and hematologic evaluation for eosinophilia are of little value in establishing the diagnosis of DUSN.
   e. In the early stages of the disease, small crops of evanescent gray-white lesions at the level of the outer retina are found in only one sector of the fundus and serve as a marker as to the location of the nematode, which is always within the vicinity of the lesions. The lesions usually fade within 7 to 10 days only to recur in an adjacent area or distant site as the nematode travels elsewhere in the eye. There may be associated mild-to-moderate vitritis and mild papillitis.
   f. In the late stages, there is optic atrophy, attenuation of the retinal vessels, and diffuse RPE degeneration.
   g. Electretinogram (ERG) is usually abnormal even when tested early in the disease course. However, it is important to note that the ERG is rarely extinguished completely in DUSN. Both cone and rod functions are affected with the b-wave affected more than the a-wave.

2. Expound on unique features
   a. The nematode found in our patient measured approximately 1733 um in length. Based on the nematode size and the patient's history of exposure to raccoons, *Baylisascaris procyonis* was the likely culprit in our case. We believe our patient
might have been infected with the *Baylisascaris procyonis* parasite during his contact with dust particles likely contaminated with raccoon feces while working at the demolition site. Saffra et al. have reported a similar case of DUSN caused by *Baylisascaris procyonis* in New York City.

V. Treatment, management

**1. Treatment and response to treatment**
   a. Direct laser photocoagulation of the nematode is the treatment of choice when the nematode can be visualized. Laser photocoagulation of the nematode was performed successfully in our patient after its localization on fundus photography. The patient was also started on an antihelminthic medication, oral albendazole 400mg twice a day for 30 days.
   b. The effectiveness of oral antihelminthic medication is controversial due its low ocular penetrance, but it may be the only treatment option when the nematode cannot be visualized. Studies by Gass et al. reported that thiabendazole might be effective in patients with DUSN when significant vitritis associated with a breakdown of blood-retinal barrier is present. Gass et al. also suggested the use of scatter laser photocoagulation in the vicinity of the active lesions to break down the blood-retinal barrier prior to the administration of thiabendazole. Although, thiabendazole is a potent antihelminthic drug, it produces serious sides with nausea, vomiting and diarrhea being the most common. Treatment with albendazole is a better-tolerated alternative. Liver function test should be obtained prior to initiating treatment due to the risk of hepatotoxicity.
   c. In conjunction with the antihelminthic agent, the patient was placed on oral prednisone 40mg daily for 2 weeks and prednisolone acetate 1% ophthalmic suspension every 2 hours in the right eye to prevent exacerbation of inflammation caused by toxins released from the destroyed nematode. Oral ranitidine 150mg twice a day was added to the regimen to prevent GI upset as a result of steroid use. A consult was ordered for the patient to be seen in infectious disease unit.

VI. Conclusion

**1. Clinical pearls, take away points if indicated**
   a. The gold standard in making the diagnosis of DUSN is the identification of the nematode
   b. Repeated examinations may be needed to find the nematode
   c. Serial fundus photography may be useful for detection of the nematode
   d. Early recognition of DUSN and prompt laser photocoagulation of the nematode are essential for preservation of vision
   e. Oral antihelminthic medication may be an effective treatment option in patients with DUSN when the nematode cannot be visualized especially if significant vitritis present.

References: