Unilateral Retinal Degeneration in a Young Hispanic Patient

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We discuss the clinical presentation, appropriate testing, differential diagnoses and management of a young Hispanic male with unilateral retinal pigment degeneration.

I. Case History

A Fourteen-year-old Hispanic male presented with reduced vision OS ongoing for 2-3 years. The patient reported a history of bumping into objects, typically on his left side. The patient reported no diplopia, headaches, eye injury/trauma, flashes of light or floaters. Medical history was unremarkable. Family history was positive for keratoconus (sister) and nyctalopia (maternal uncle) of unknown etiology. The patient was born in a small town in Mexico, and migrated to the United States as an infant. Since then, the patient has resided in New York City.

II. Pertinent Findings

Entering acuities without correction were 20/20 OD and 20/400 OS. Near acuities without correction were 20/20 OD and 20/100 OS. Pupils demonstrated a left grade II afferent pupillary defect. Confrontational field were full to finger counting OD and constricted in all quadrants OS. Ishihara color vision plates were reduced in both eyes; 10/14 OD and 1/14 (test plate only) OS. Brightness comparison was reduced OS by 70%. The patient had no stereopsis. Refraction revealed emmetropia OD and compound myopic astigmatism OS (-3.75-1.00x150), yielding best-corrected acuities of 20/20 OD, 20/40 OS. Dilated fundus examination revealed a pink and distinct optic nerve with mid-peripheral pigmentary changes superior-temporally in the right eye. The left eye revealed a pink tilted nerve surrounded by 360 degrees of peripapillary atrophy with scattered intra-retinal, white, punctate dots in the posterior pole and mid-peripheral subretinal pigmented scars superiorly, temporally and nasally. The left eye showed no sign of active inflammation.

Additional color vision testing using the Farnesworth Dichromotous Test Panel D15 demonstrated no color defects. The left eye demonstrated color defects without specificity. Corneal topography revealed no signs of keratoconus. Automated 30 degree visual field testing revealed no defects in the right eye and a constricted field to 15-20 degrees in the left eye with reduced sensitivity within the central 15 degrees. Goldmann visual fields revealed normal sensitivity in the temporal field but constriction elsewhere to the central 20 degrees in the left eye. Optical coherence tomography (OCT) revealed an intact photoreceptor integrity line without abnormalities OD. The OCT OS demonstrated a disrupted photoreceptor integrity line that was completely absent in and outside the macula. A full-field electroretinogram revealed normal scotopic, photopic and flicker (cone) responses in the right eye. The ERG in the left eye was attenuated under all conditions. Both accompanying family members (mother and half-sister) agreed to examinations with dilation at the time of these discoveries and both were free of pathology.

III. Differential Diagnosis
Based upon the patient’s symptoms and clinical findings differential diagnoses included diffuse unilateral subacute neuroretinitis, unilateral retinitis pigmentosa and ophthalmic artery occlusion.

IV. Diagnosis and discussion

The differentials of a unilateral RP-like fundus are discussed below and include the following:

1. Diffuse unilateral subacute neuroretinitis (DUSN) is an inflammatory condition caused by a nematode found in the subretinal space. One identified nematode is *Ancylostoma caninum*, most frequently found in the Southeastern United States, the Caribbean, and Latin America. The second is *Baylisascaris procyonis*, which has been identified in northern and Midwestern United States and parts of Europe amongst the raccoon populations\(^1\). A motile worm found on fundus photography was identified as *Baylisascaris procyonis* based on morphologic evaluation in a fifteen year old female from Brooklyn, New York. She presented with unilateral painless vision loss with diffuse retinal pigment changes, arteriole attenuation and optic nerve pallor\(^2\). Additionally, a case of *Baylisascaris procyonis* neural larva migrans in an infant, also in the boroughs of New York City has been identified, alerting pediatricians in the local health community\(^3\). Gass et al\(^4\) describe DUSN as widespread diffuse and focal pigment epithelium changes sparing the macula, peripheral visual loss, attenuated arterioles and optic atrophy. Gass and Braunstein\(^5\) reported decreased electroretinogram (ERG) responses in cases of DUSN, but are rarely extinguished.

2. Ophthalmic artery occlusions can mimic retinal changes similar to that of unilateral RP. This diagnosis is usually supported by a history of vision loss immediately following face mask anesthesia, which is the more common etiology or neurosurgical procedures, for intracranial aneurysms for example, that would compromise the ophthalmic artery circulation. Reduced perfusion of the ophthalmic artery causes retinal edema, retinal arteriolar narrowing and a pale optic disc, resulting in severe vision loss\(^6\).

3. Unilateral retinitis pigmentosa (RP) is a controversial diagnosis, and only a handful of cases have ever been reported\(^5\), if they are, in fact, unilateral RP. Diffuse RP is a bilateral usually progressive disease characterized by pigmentary retinopathy (bone-spicule pigmentation along the blood vessels), attenuated blood vessels, waxy pale optic nerve and reduced peripheral field as the disease progresses. Nyctalopia is the primary complaint and the visual field is constricted in all meridians. Unilateral forms of RP would be a diagnosis of exclusion and would require genetic testing for additional support of the diagnosis as well as family history.

Based on our patient’s history and examination results, we believe he most likely has DUSN. Although the infection occurred years ago and a live worm was not found on examination, the visual field in the affected eye is preserved in the temporal field and the other eye does not demonstrate any structural or functional evidence of RP.

V. Treatment and management

Our patient is current being monitored every 2-3 months for changes in visual acuity, color vision, automated visual field and fundus examinations. In the event a nematode is identified the worm is commonly treated with photocoagulation. Systemic steroids can supplement treatment to control the inflammatory response. In order to determine if an underlying systemic condition exists, further
ancillary testing, such as *Baylisascaris procyonis* larval excretory–secretory antigens by enzyme-linked immunosorbent assay, is also being considered.

VI. Conclusion

This is a case report of a unilateral RP-like degeneration, supported by fundus examination and ERG findings, but which is most likely DUSN, related to infection involving a roundworm or nematode organism. It is not expected to progress over time, unlike RP, but these patients must be monitored at the very least every 6 months for changes.
References: