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Case report

Optical coherence tomography angiography of diffuse unilateral subacute neuroretinitis

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ABSTRACT

Purpose: Diffuse unilateral subacute neuroretinitis (DUSN) is often a challenging diagnosis to make. We present a DUSN case with its multimodal imaging to aid in the diagnosis, emphasizing the observations on optical coherence tomography angiography (OCTA).
Observations: The evolution of a DUSN case is presented. Fundus photography and OCTA aided in the identification of the nematode.
Conclusions and importance: DUSN is a difficult diagnosis to establish. We report the first case to our knowledge in which OCTA aided in the diagnosis of DUSN.

1. Introduction

Diffuse unilateral subacute neuroretinitis (DUSN) was initially described by Gass[1] in 1978. This paper came fifteen years after he and his colleagues recognized a "unilateral retinal wipe-out syndrome" in which healthy young individuals developed: "(1) insidious, usually severe loss of peripheral and central vision; (2) vitritis; (3) diffuse and focal pigment epithelial derangement with relative sparing of the macula; (4) narrowing of the retinal vessels; (5) optic atrophy; (6) increased retinal circulation time; and (7) subnormal electroretinographic findings." (Gass and Scelfo).[1–4] Patients can present in the early stage[5] when the predominant findings are of chorioretinitis or in the late stage[6] with poor vision, retinal and optic atrophy.

Most DUSN patients are young, present with altered vision and are in the late stages of the disease. Various nematode etiologies have been proposed depending on the size, speed of the nematode and the geographic area in which it was contracted.[2–4,7] Most commonly, Baylisascaris procyonis and Ancylostoma caninum are thought to be the culprit. Precise diagnosis of the nematode is limited due to lack of good serologic testing or stool examinations and most reports are made based on clinical exam. Specifically, that B. procyonis is longer and thought to be greater than 2000 μm while A. caninum is less between 250 and 500 μm.[2–4]

Use of a contact lens to thoroughly examine the retina or examining color fundus photos have been generally recommended to help locate the worm. Despite these efforts and a strong clinical suspicion for DUSN, visualization of the worm on examination is uncommon and seen in only 25–40% of cases.[2–4] OCT or OCTA have not been previously described in the identification of the worm in cases of DUSN. Laser photocoagulation applied to an identified worm is desired to limit retinal damage. Oral antihelminthics can be used in addition to laser to treat systemically[8] or if a worm is not identified and the suspicion for DUSN is high.

2. Case report

A 49-year-old man presented with significantly decreased vision in the right eye for 2 months. He was previously treated with lisinopril for hypertension and, at presentation to our clinic, was on 80 mg prednisone for intraocular inflammation. He was a tree-trimmer by occupation in Northern California. Review of

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systems revealed he had experienced widespread urticaria 1 month prior to presentation. No family history of ocular problems.

Visual acuity was hand motion (HM) and 20/16 in his right and left eye, respectively. A relative afferent papillary defect was present in the right eye. Intraocular pressure was within normal limits. The anterior segment examination was unremarkable. A posterior segment examination of the right eye revealed moderate vitritis, temporal optic disc pallor with severely attenuated retinal vessels (Fig. 1). In the macula, a motile subretinal worm (2500 μm x 100 μm) was identified with active retinitis seen superiorly (Fig. 2). Subretinal fibrosis and widespread retinal pigment epithelium (RPE) atrophy was also present. Serial fundus photography revealed significant progression from his exam 1 month earlier (Fig. 3). Posterior segment examination of the left eye was unremarkable.

Spectral-domain optical coherence tomography (SD-OCT) of the right eye revealed diffuse atrophy of both the inner and outer retinal layers as well as the RPE (Fig. 4). Prominent internal limiting membrane (ILM) was present (Oréfice's sign). Wide-field fundus autofluorescence of the right eye showed diffuse, alternating, speckled hyper- and hypo-autofluorescence predominantly in the posterior pole (Fig. 5). Wide-field fluorescein angiography of the right eye revealed transmission hyperfluorescence in the areas of atrophy corresponding with window defect (Fig. 6). OCT angiography (OCTA) was performed on this patient and fortuitously, it revealed what we believed to be an anomalous pattern in the superotemporal macula (Fig. 7). As seen from the color fundus photos performed a few moments later, OCTA in fact revealed the mobile worm. The caliber and configuration seen on OCTA is consistent with a nematode. The cross-section OCTA reveals the worm within the retina curled upon itself (Fig. 8). Once the worm had moved again, OCTA was re-performed and it was confirmed that only normal retinal vasculature was seen and the anomalous pattern had disappeared. Laser photocoagulation was applied to the worm along its entire body. A 4-week course of oral albendazole was started and oral prednisone was tapered.

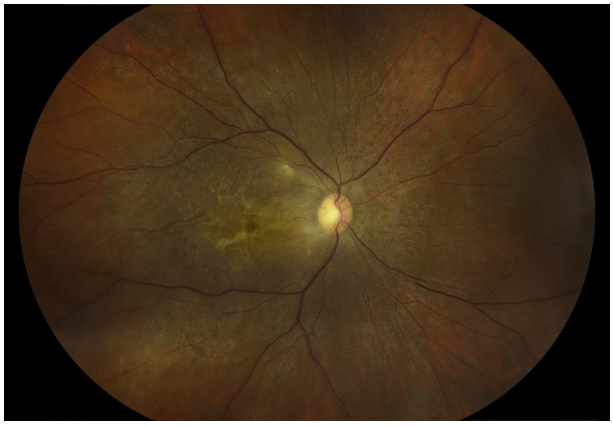

Fig. 1. Wide-field color fundus photo of the right eye revealing diffuse retinal pigment epithelium atrophy, fibrosis, attenuated vessels and optic disc pallor. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

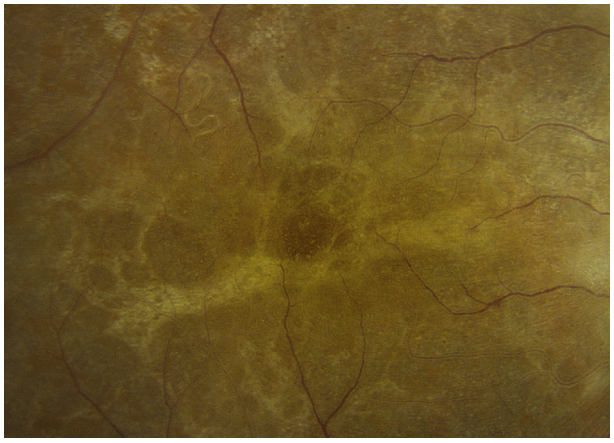

Fig. 2. Color fundus photo of the right macula revealing a worm. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

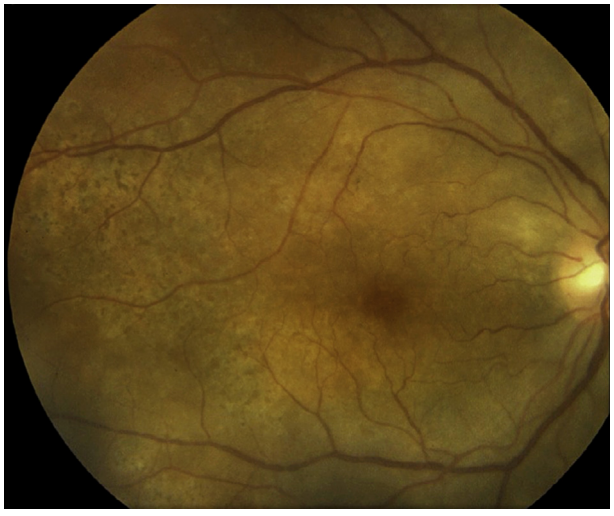

Fig. 3. Color fundus photo of the right eye 1 month prior with what appears to be widespread retinal whitening. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

3. Discussion

Diffuse unilateral subacute neuroretinitis is an uncommon diagnosis. The prevalence of DUSN may actually be higher than reported due to the challenging course of the disease and the difficulty identifying the nematode. For example, there is a growing belief that many patients previously diagnosed with unilateral retinitis pigmentosa are in fact cases of undiagnosed DUSN.

Gass who initially described DUSN felt that high quality color fundus photos was the best way to identify nematodes. Our case proves that fundus photos still remain a valuable method of identification. However, with the worm not found in over 50% of suspected DUSN cases, there is still significant room for other techniques. To the best of our knowledge, this is the first report of OCTA aiding in the diagnosis of DUSN. Our case demonstrates

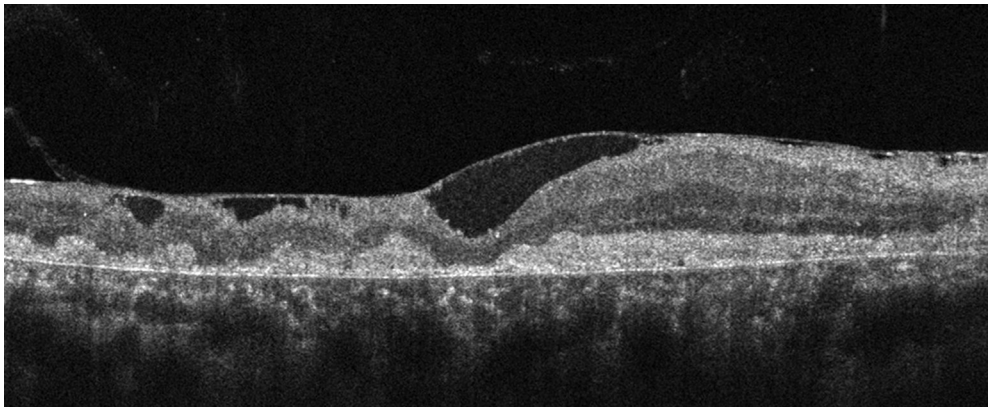

Fig. 4. Spectral-domain optical coherence tomography imaging of the right eye revealing diffuse retinal and retinal pigment epithelium atrophy.

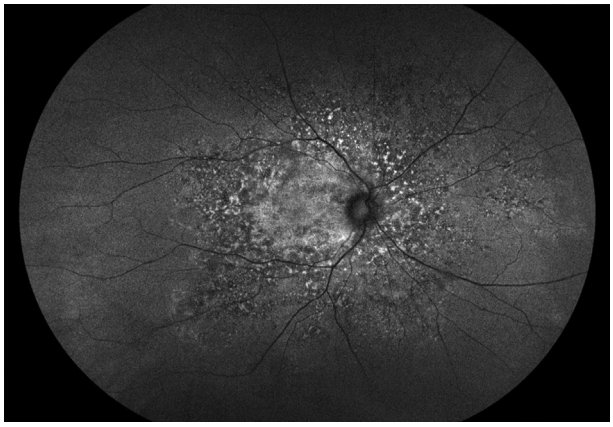

Fig. 5. Wide-field fundus autofluorescence of the right eye revealing diffuse speckled hyper and hypoautofluorescence.

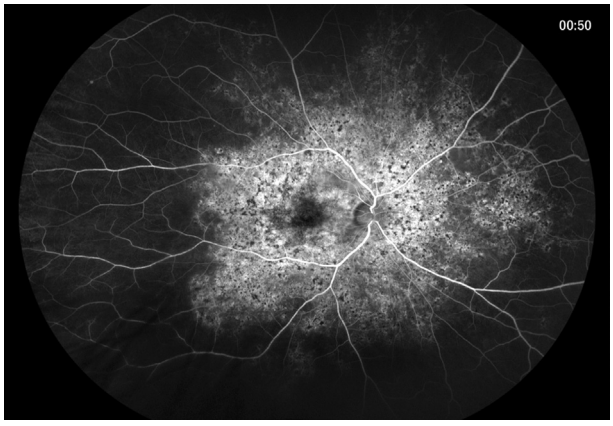

Fig. 6. Wide-field fluorescein angiography of the right eye revealed widespread hyperfluorescence.

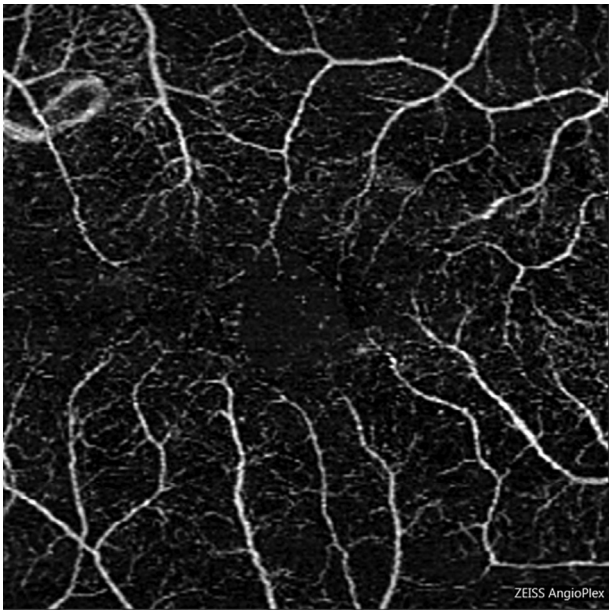

Fig. 7. Optical coherence tomography angiography (en-face) of the right eye revealed a mobile intraretinal worm.

that OCTA is another imaging modality that can diagnose DUSN. The reason that the nematode was detected is likely due to movement of the worm as nematodes have no vascular system. Thus, it is possible that an inactive worm may not be detected.

The addition of OCTA to the widely accepted diagnostic tests performed for suspected DUSN cases may help increase the rate of diagnosis. Specifically, in the acute phase when it is most difficult to diagnose it as it is masquerading as a chorioretinitis, and even more so if the causative agent is the smaller Ancylostoma caninum larva; OCTA may help recognize it earlier and consequently save critical vision. OCTA may also provide further understanding to the disease process as we will be able to pinpoint the location of the worm, be it subretinal, intraretinal or within the choroid. As higher resolution and ultra-widefield OCTA becomes available, our capability to diagnose and understand DUSN will likely improve.

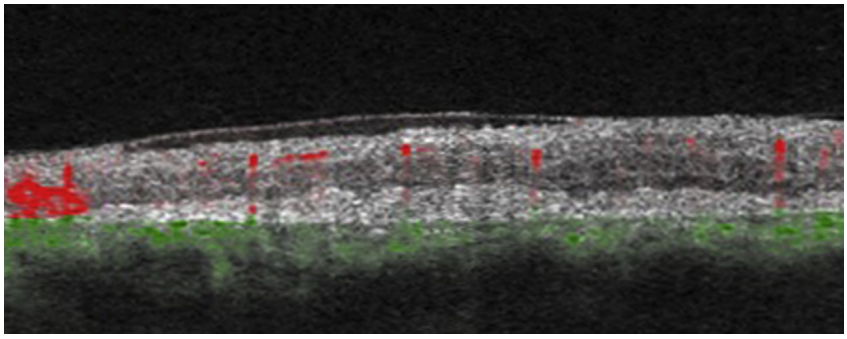

Fig. 8. Optical coherence tomography angiography (cross-section) of the right eye revealed a mobile intraretinal worm.

Patient consent

We further confirm that any aspect of the work covered in this manuscript has been conducted with the ethical approval of all relevant bodies. Consent was obtained from the patient in writing.

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Conflict of interest

The following authors have no financial disclosures: AK, MJ.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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