Diffuse Unilateral Subacute Neuroretinitis
A Case Report

Sony Siraj E DO, Reena A MS DO, Thomas George MS, DO

Donald M. Gass and his colleagues recognized a "unilateral wipe out syndrome" in which healthy young individuals developed insidious usually severe loss of peripheral and central vision, vitritis, diffuse and focal PED with relative sparing of macula, narrowing of retinal vessels, optic atrophy, increased retinal circulation time and subretinal electroretinographic findings. Later it was called as Diffuse Unilateral Subacute Neuroretinitis. We report here an interesting case of a 10 year old boy who was suspected to have DUSN and his condition completely resolved after a course of antihelminthic therapy.

Key Words: Dusn, Ped, Mewds, Apmppe.

Case Report

A 10 year old boy presented to us at Regional Institute of Ophthalmology, OPD with complaints of defective vision (RE) of 2 weeks duration. He was on treatment for hydronephrosis and protein energy malnutrition. He had the habit of taking non-vegetarian diet frequently from hotel and gave a history of deworming every 3 months. General examination revealed no relevant findings. Ocular examination showed a BCVA of 6/6 (RE) and 6/12 (LE). Anterior segment was within normal limits. Fundus of the LE showed a hyperemic disc with a lobulated cystic subretinal lesion of size 2DD beneath the superotemporal arcade, 1DD away from the superior border of the disc (Fig. 1). Superotemporal arcade vessels showed sheathing. Macular edema along with macular star was seen. Multiple subretinal patches were there in the superotemporal quadrant, 1DD temporal to the cystic lesion, suggestive of track lesion. A provisional diagnosis of DUSN was made. Fundus of the RE was within normal limits.

Blood investigations showed a total count of 9600 per cumm and a differential count of P49 (polymorphs) L46 (Lymphocytes) E5 (Eosinophils). CRP (C-Reactive protein) value was raised (1.2mg %). B scan revealed retinochoroidal complex thickening and oedema of the optic nerve. Patient was started on Tab Albendazole 400 mg od and after 24 hrs systemic steroids were given. Patient showed dramatic improvement on day 3 of treatment. After 1month of treatment patient had a BCVA of 6/6 (BE). Fundus picture showed a complete resolution of the subretinal lesion and macular star.

Fig. 1. Fundus pictures of left eye. Note the hyperemic disc, subretinal lobulated cystic swelling, macular star and suggestion of a ‘track-sign’.
Systemic steroids were tapered over 1 month and albendazole therapy was also stopped after 1 month.

Discussion

DUSN, also called as the unilateral retinal wipe out syndrome typically affects children and young adults. Several species of nematodes, including Toxocara canis, Baylisascaris procyonis, and Ancylostoma caninum have been suggested as the potential etiologic agent of DUSN. The nematodes have been classified into 2 different sizes. The smaller nematode, measuring 400 to 1000 μm in length, is endemic to the southeastern United States, the Caribbean islands, and Brazil. The larger nematode, measuring 1500 to 2000 μm in length, has been described in the northern midwestern United States. Usually there are no associated systemic symptoms although cutaneous and neural larva migrans have been described in a few patients. The proposed mechanism of vision loss include the host inflammatory reaction to the parasite, toxic effect of the worm’s secretary proteins, mechanical damage produced by the movement of the worm or an autoimmune reaction somehow initiated by the infection. The clinical features of DUSN manifest as early and late stages. In the early stages (vision 6/6 – 6/60), the external and slit lamp examination is often normal. Early features include retinal arteriolar narrowing, intraretinal perivascularexudates, pigment epithelial depigmentation and recurrent multifocal evanescent grey white lesion in the outer retina. The retinitis is found typically in one sector of the fundus and can provide a clue to the worm location. The retinitis resolves in 7 to 10 days with minimal or no residual retinal changes. Occasionally the worm is identified with fundus photography.

In the late stages, vision is typically 6/60 or less with a dense central scotoma. Optic atrophy and vascular attenuation are prominent features. A subretinal mass associated with choroidal neovascularization has been described in the macula and around the optic nerve. ERG shows abnormal rod and cone function in the affected eye with a reduction of b/a wave amplitude ratio suggestive of inner retinal injury. Eosinophilia is rare in DUSN. Macular cyst has been reported to be associated with DUSN as an interesting and unusual finding. In patients with diffuse unilateral subacute neuroretinitis (DUSN), the presence and, therefore, clinical visualization of subretinal nematode makes the diagnosis obvious. However when located under the retinal pigment epithelium (RPE), diagnosis is presumptive and challenging. The appearance of sub-RPE serpiginous tract, peripheral RPE hypopigmentation and good clinical response to anti-helminthics support the diagnosis. Arundhati Anshu and Soon Phaik Chee published a case report of presumed DUSN, where subretinal live worm was not seen and the patient responded well to anti-helminthic therapy. So it is important to have a high index of suspicion when patients present with a combination of above findings. This will help in early control of ocular inflammation and also in salvaging vision. Other rare presentations include a case report of diffuse unilateral subacute neuroretinitis (DUSN) that developed an acute iridocyclitis with hypopyon after a year of follow-up and resolved after treatment with systemic corticosteroid. (Cristina Muccioli et al).

Differential diagnosis to be considered include intermediate uveitis, Pars planitis, MEWDS, APMPPE, toxoplasmosis, sarcoidosis, syphilis, Behcet’s disease, atypical RP, siderosis etc.

Laser photoocoagulation (Xenon or Argon) should be considered as the first line of therapy in patients in whom motile larva are identified provided the treatment will spare the macula. The role of a combination of laser treatment, systemic steroid, and antihelminthics is also proposed. For the 50% of patients in whom a worm cannot be found, a month long course of albendazole (400 mg od) along with
systemic steroids should be considered. Immobilization of the subretinal nematode has been observed following systemic antihelminthic therapy, and so it has been recommended that patients with DUSN in whom worm cannot be initially identified receive a course of such therapy in order to maximize the chances of identifying and treating the offending organism.

References