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Case Report

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Neuroretinitis: a case report of a 16-year old high school student

Abstract

A 16-year old senior high school girl presented with painless progressive blurring of vision in the right eye at Sight for Africa Eye Clinic in Darkuman, Ghana. The condition, she reported, started 14 days ago. There was no history of ocular trauma, refractive error, or any sight-threatening condition such as glaucoma. Her medical history, as far as she was concerned, was negative. She had not come into contact with cats or other pets recently or in the past. Following a detailed history taking and a comprehensive examination, she was diagnosed with idiopathic neuroretinitis. Neuroretinitis is a focal inflammation of the optic nerve and peripapillary retina or macula of either infectious or idiopathic etiology, and is characterized by acute unilateral vision loss and macular star-shaped exudates.

Keywords: neuroretinitis, visual loss, optic neuritis, unilateral, macular star

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Introduction

Neuroretinitis is a focal inflammation of the optic nerve and peripapillary retina or macula. It can be either infectious or idiopathic and is characterized by acute unilateral vision loss.[1,2] Leber originally described it in 1916 as a "stellate maculopathy"[3] but Don Gass disputed this definition in 1977, citing that disc edema precedes macular exudates.[4] Consequently, Gass established optic disc leakage by fluorescein angiography and suggested the term "neuroretinitis". Neuroretinitis, is thus, one form of optic neuritis, though it is rarely reported comparative to other forms such as retrobulbar neuritis and papillitis. Funduscopically, neuroretinitis is often confused with hypertensive, renal, and infiltrative retinopathies as well as with papillitis, papilledema, anterior ischaemic optic neuropathy, and retinal vein occlusion.[5] The pathophysiology of neuroretinitis is characterized by an inflammation of the optic disc vasculature with exudation of fluid into the peripapillary retina.[2] The lipid-rich component of the exudate is further able to penetrate into the outer plexiform layer, forming a 'macular star' pattern. Only the aqueous phase is then able to pass through the external limiting membrane to accumulate beneath the neurosensory retina.

Based on the etiology and relapse, neuroretinitis can be classified into the following categories. For the first category, a specific infectious agent is implicated. Commonest among the infectious causes is cat-scratch disease while Rocky Mountain spotted fever, toxoplasmosis, toxocariasis, histoplasmosis, leptospirosis, syphilis and Lyme disease are the rare infectious causes. In this infectious type, broad-spectrum antibiotics like azithromycin are usually prescribed.[6,7] In the second category, where there is no definite infectious or inflammatory cause, the condition is termed idiopathic neuroretinitis or Leber's idiopathic stellate retinopathy.[3] It has been reported that visual prognosis in patients with idiopathic neuroretinitis is excellent, with or without interventions. Albeit no well-defined treatment plan exist, the use of corticosteroids has proven beneficial in some reported studies.[8] The third category, which is termed recurrent idiopathic neuroretinitis, is a variant of idiopathic neuroretinitis where there is relapse of the condition. Visual loss in this case takes the form of an optic disc disease and visual defects are central and nerve fiber layer-related. For recurrent idiopathic neuroretinitis, long-term immunosuppressant is usually considered.[9]

Case report

On Tuesday, the 26th of January 2016, at about 11:00am, a 16-year old senior high school girl presented with painless progressive blurring of vision in the right eye at Sight for Africa (SFA) Eye Clinic, Darkuman. The condition, she reported, started 14 days ago. There was no history of ocular trauma, refractive error or any sight-threatening condition such as glaucoma. Her medical history, as far as she was concerned, was negative. She had not come into contact with cats or other pets recently or in the past. She reported of no symptoms of fever, fatigue, flu, etc., at the onset of this problem. But for a pair of reading glasses worn by her father, her parents' ocular history was nil. Her mother was diagnosed with diabetes a couple of years ago. She was not on any medication prior to the onset of this condition and there were no known drug allergies or environmental allergens. On examination, a relative afferent pupillary defect with best-corrected visual acuity of 20/80 in the right eye and 20/20 in the left eye was noted. A slit-lamp examination disclosed that the anterior segment was unremarkable in the right eye. Fundus examination of the right eye revealed an optic disc with indistinct margins, active retinal lesions, disc lesions, disc oedema and star-shaped exudates distributed in the macula. There was also deep subretinal exudation along the superotemporal vessels. The retinal periphery, however, was normal in the right eye. The ability to perceive colour was impaired in the right eye. The left eye was unremarkable in all aspects.

Figure 1 Macular star pattern of neuroretinitis of the right eye.

Ocular examination

A Snellen Visual Acuity chart was used to determine patient's visual acuity and subjective refraction was done via a trial lens set. A non-contact tonometer was used to determine the intraocular pressures and an ophthalmoscope was used to examine both anterior and posterior segments of the eye.

i. Visual acuity(uncorrected)

20/200 OD

20/20 OS

ii. Subjective refraction

OD -1.50 Dioptres 20/80

OS Plano 20/20

iii. Pupils

The left eye was reactive to light. Relative Afferent Pupillary Defect (RAPD) was seen in the right eye.

iv. Extraocular motility

Smooth, Accurate, Full and Extensive (SAFE)

v. Intra-ocular pressure

11mmHg OD, 12mmHg OS

vi. External and anterior segment examination

Normal OU

vii. Fundoscopy

OD: clear media with few vitreous cells, marked optic disc oedema, active retinal and disc lesions, exudation of fluid from the optic nerve into the macula, inflamed vessels, no loss of retinal nerve fibre layer (RNFL), normal periphery. OS: Transparent ocular media, no retinal nerve fibre layer (RNFL) loss, normal macula and vessels, normal periphery.

Where

OD represents right eye

OS represents left eye

OU represents both eyes

RAPD represents Relative Afferent Pupillary Defect.

Differential diagnoses

The following were the differential diagnoses for this case:

I. Papillitis

II. Papilledema

III. Anterior Ischaemic Optic Neuropathy (AION)

IV. Central Retinal Vein Occlusion (CRVO)

Diagnosis

A diagnosis of neuroretinitis was made based on the clinical presentation. Thus, the marked reduction in visual acuity, the appearance of the right optic nerve, the star-shaped macula, as well as the accumulation of fluid in the retina is a depiction of this condition.

Plan of management

The patient was prescribed oral prednisolone 10 milligrams to be taken for 10 days and she was requested to perform the following tests and then report in a week's time.

I. Fasting blood sugar, blood pressure and other serological tests.

II. Magnetic Resonance Imaging (MRI) and Computed Tomography (CT)

III. Fundus Photography

IV. Visual Field Test (VFT)

First review (on second visit)

Patient looks cheerful and reported that her vision in the right eye had improved considerably.

Ocular examination

i. Visual acuity (uncorrected)

$20/40^{2}$-OD

20/20 OS

ii. Pupils

Left eye was reactive to light. Relative Afferent Pupillary Defect (RAPD) was seen in the right eye.

iii. Extraocular motility

Smooth, Accurate, Full and Extensive (SAFE)

iv. Intra-ocular pressure

22 mmHg OD, 23 mmHg OS

v. External and anterior segment examination

Normal OU

vi. Fundoscopy

OD: Normal media, improved optic disc oedema, resolution of retinal and disc lesions, resolution of subretinal and intraretinal fluid in the macula, resolution of exudation along the superotemporal vessels, normal periphery.

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OS: Transparent ocular media, pseudopapillitis, no loss of retinal nerve fibre layer (RNFL), normal macula and vessels, normal periphery.

Results of laboratory and clinical investigations

Patient blood glucose as well as blood pressure was normal. Visual field testing revealed a centrocaecal scotoma in the right eye whereas the left eye fields were normal. Radiological examination of the skull and paranasal sinuses was also normal. The results of all blood tests, including serological tests for syphilis, Lyme disease, toxoplasmosis and toxocariasis proved negative. ELISA for cysticercosis, tuberculosis, Lyme disease, leptospirosis, brucellosis, toxocariasis and toxoplasmosis and Weil-Felix test showed negative result. Findings from CT and MRI scan of the brain were also devoid of any abnormality.

Diagnosis

Based on the findings above, it was finally established that the patient had idiopathic neuroretinitis.

Management plan

Patient was prescribed oral prednisolone 5milligrams to be taken for 10 days and a second review was scheduled in 14 days.

Discussion

Neuroretinitis is a clinical diagnosis made based on a number of ocular findings in addition to a comprehensive history taking. These signs following a unilateral painless vision loss are optic disc oedema, macular oedema and a later development of a macular sign. In the acute phase, vitreal cells are commonly seen. The condition affects persons of all ages and often more common in the third and fourth decade of life, with no gender preference. In the absence of a proven etiology to the disease, the condition is diagnosed as Leber's idiopathic stellate retinopathy or idiopathic neuroretinitis. It is a diagnosis of exclusion made after ruling out other known causes of neuroretinitis. It occurs most in healthy young subjects presenting with acute unilateral vision loss.

Visual acuity at the time of initial examination ranges from 20/30 to 20/200. The most common field defect is caecocentral scotoma, but central scotomas, arcuate defects and even altitudinal defects may also be present. Unless the condition is bilateral, Relative Afferent Pupillary Defect (RAPD) is present in most patients. The degree of the optic disc swelling may be mild, moderate or severe depending in part, on the time of the first examination. Flame shaped haemorrhages may be present in very severe cases. A macular star-shaped composed of lipid (hard exudates) may not be present when the patient is examined immediately after the onset of visual symptoms, but tends to become prominent as the optic disc swelling resolves. An early colour vision defect is not uncommon as occurs in any optic nerve dysfunction.

Conclusion

Idiopathic neuroretinitis is self-limiting but patients are often treated with systemic steroids in the acute phase. In this idiopathic variant, a general prognosis for visual recovery is good, as was the case with this patient. Patient here responded very well to the steroids as seen in the elevation of her intraocular pressure on her second visit. In the secondary forms wherein there is an identified or strongly associated infectious agent, specific therapy against the organism along with steroids appears justifiable from anecdotal reports. It is worth noting that neuroretinitis may be easily confused with other clinical entities such as papillitis, papillledema, central retinal vein occlusion (CRVO) and anterior ischaemic optic neuropathy (AION), amongst others. Hence a very detailed history taking together with a thorough investigation should be carried out.

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Conflicts of interest

There are no conflicts of interest.

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