

Case Series

INITIAL DIAGNOSTIC DILEMMA WITH 3 NEURO-OPHTHALMIC CASES SEEN AT UNIVERSITY OF PORT HARCOURT TEACHING HOSPITAL, NIGERIA

ABSTRACT

AIM: The aim is to present three patients with neuro-ophthalmic symptoms and signs who initially presented a diagnostic dilemma. Two later turned out to be retroviral positive.

PRESENTATION OF CASES: CASE 1 was a 35 yo female nurse assistant who first presented with a one month history of deep-seated pain in the right eye, right side of face and headache with sudden onset diminished vision in the right eye. She was initially diagnosed with optic neuritis but this was followed 7 months later with panuveitis and bullous RD. Patient was not hypertensive or diabetic and was retroviral (RV) negative. Even though she presented a diagnostic dilemma, a tentative diagnosis of recurrent idiopathic neuro-retinitis is entertained on account of the clinical features and normal investigations. CASE 2 was a 32yo female who presented on account of sudden onset binocular horizontal diplopia of 2 weeks duration. She later developed dizziness and paraesthesia of the left side of her body and examination revealed bilateral torsional nystagmus. The patient was HIV positive with a CD4⁺=39 cells/μl. The third Case was a 17yo student initially diagnosed with Bell's palsy but later developed transient visual loss. He was found to be HIV positive with a CD4⁺= 768 cells/μl.

CONCLUSION: These 3 cases bring to the fore the importance of entertaining a high index of suspicion in managing unusual cases that present to the ophthalmologist; and in all cases to rule-out HIV/AIDS since it has its highest prevalence in sub-Saharan Africa. That way, early intervention can be instituted and this would ultimately reduce morbidity and mortality from the disease.

KEY WORDS: HIV/AIDS, Neuro-ophthalmic presentation, Diagnostic Dilemma

INTRODUCTION

The human immunodeficiency virus (HIV) infection still poses a major health challenge in many developing countries including Nigeria. Infection by HIV involves almost all the systems in body and neuro-ophthalmic problems are known to occur both in human immunodeficiency virus (HIV) infection and AIDS. Central nervous system (CNS) opportunistic infections and malignancies such as lymphoma are the major source of these complications; but some however result from direct effect of the virus on the CNS.¹ Neuro-ophthalmic signs may sometimes be the initial manifestation of AIDS,¹ and the nervous system is involved in up to 50% of HIV infected subjects;²⁻⁴ with histological studies showing that in up to 75–90% of AIDS patients, damage to the brain and optic nerve can occur.⁵ Neuro-ophthalmic disturbances have been described in both asymptomatic HIV positive patients and in those with full blown AIDS. In patients with AIDS there is a 3%–8% incidence of neuro-ophthalmic disorders.⁶⁻⁸ In a study at the Infectious Diseases Unit of Kinshasa University Hospital, neuro-ophthalmic manifestations were noted in 60% of their patients with ocular movement abnormalities being the most common followed by visual field defects, optic neuropathy, papilledema and ocular cranial nerve palsies.⁹ In their study in Ethiopia, Bekele et al. reported only about 3.2% cases of neuro-ophthalmic complications in patients who were on HAART therapy.¹⁰

The aim of this study is to report three cases of patients who had neuro-ophthalmic symptoms and signs that initially presented a diagnostic dilemma. Two of the cases later turned out to be retroviral sero-positive but for the first patient, the diagnosis is still unknown but may likely be a case of recurrent idiopathic neuro-retinitis in the same eye.

PRESENTATION OF CASES

CASE 1

FW is a 35 yo female nurse assistant who first presented at our Clinic in August 2017 on account of a one-month history of deep-seated pain in the right eye, right-sided facial pain and right-sided headache associated with sudden onset diminished vision in the right eye. Patient had severe photophobia but there was no pain on eye movement. Patient was not a known hypertensive or diabetic.

At presentation, unaided visual acuity in the right eye (RE) was **Count Fingers (CF) @ 3M** and this improved with pinhole to 6/18. Visual acuity in the left eye was 6/6. The

eyelids, conjunctiva, cornea and anterior chamber were all normal. Other examination results were a relative afferent pupillary defect (RAPD) Grade II and disc edema in the RE. There was color desaturation and patient could only correctly identify 5/11 on the Ishihara color vision plate. The left eye was normal. The intraocular pressures (IOP) were also within normal limits – 15mmHg and 12mmHg respectively and her body mass index (BMI) was 31. An initial diagnosis of Right Optic Neuritis to rule out Idiopathic Intracranial Hypertension (IIH) was made and the patient commenced on systemic steroids (IV methyl prednisolone 1gm daily for 3 days was recommended but due to unavailability, tablets 60mg daily for 14 days was used and this was followed by a short-term taper). One week later, the vision improved from CF to 6/6 in the RE and there was resolution of other symptoms except for RAPD that persisted in the RE. Two months after her initial presentation, patient came with complaints of mild right ocular pain and foreign body sensation, but visual acuity still remained normal. Apart from an elevated erythrocyte sedimentation rate (ESR) (31mm/hour), other investigations – full blood count (FBC), fasting blood sugar (FBS), Chest radiograph, serum angiotensin I-converting enzyme (ACE) levels and retroviral screening were all within normal limits. Seven months later, patient presented back at our Clinic on account of pain on eye movement in the RE. Ocular examination revealed that her VA had dropped to 6/36 from 6/6; there was RAPD grade III, disc edema and superior half macular star in the RE. (Fig. 1) Patient was then re-commenced on systemic steroids and fasting lipid profile (FLP) and central visual field (CVF) test were ordered. The FLP was within normal limits but the CVF analysis showed a superior altitudinal defect. Ten months after her first presentation, the patient reported back with worsening vision in the RE, tearing and redness. Ocular examination showed that her VA had dropped further to LP in the right eye; there was ptosis, ciliary injection, aqueous flare and cells, and seclusion pupillae (Figure 2). Patient had also developed a complicated cataract associated with severe vitritis. Patient was then managed for pan-uveitis with systemic steroids, septrin and daraprim in addition to topical steroid/antibiotic combination. On follow-up one week later, patient had developed an inferior Bullous Retinal Detachment (Figure 3) in that right eye.



Figure 1: Neuro-retinitis Figure 2: Complicated cataract Figure 3: Bullous RD

CASE 2

OC is a 32yo newly married public servant who presented at our Clinic in May 2018 on account of sudden onset binocular horizontal diplopia of 2 weeks duration. There was no history of prior systemic illness such as DM or hypertension, and there were no Ear, Nose and Throat (ENT) symptoms. At presentation, patient was very ill looking with a pinhole visual acuity of 6/18 in the RE and 6/24 in the LE. There was moderate ptosis in the right and she had mixed torsional and vertical nystagmus in both eyes. Extra-ocular motility in both eyes was full range. The initial diagnosis was Acquired Nystagmus to rule out a space-occupying lesion (SOL). The following investigations were ordered – magnetic resonance imaging (MRI) of Brain and Orbits, FBS and FBC. Uni-ocular patching was recommended while awaiting investigation results and patient commenced on tablets prednisolone 20mg daily. On follow up a week later, the symptoms had begun to resolve and patient was much better. All investigation results were noted to be within normal limits.

Patient however re-presented one month later with complaints of left-sided paraesthesia, left-sided headache, tearing, and severe photophobia in the left eye, there was also a history of dizziness while walking. She complained of a generalized body rash that erupted a few weeks before presentation and said she thought the body rash was as a result of the body cream she was using at that time. On examination, patient was noted to be in severe distress even though the nystagmus had resolved. There was severe photophobia in the LE and VA was 6/24 in both eyes. She had diffuse conjunctival injection and severe left vitritis but the corneal and anterior chamber were normal. Her IOPs were normal (RE=15mmHg; LE=17mmHg). A diagnosis of Migraine without Aura was made and patient commenced on sumatriptan 100mg daily and topical steroids and

tropicamide. Retroviral test was requested for. One week later, patient came back with severe drop in vision to HM in the LE as a result of panuveitis (keratic precipitates - KP⁺⁺⁺, flare, cells and vitritis) with elevated IOPs(RE = 21mmHg and LE = 25mmHg). Patient was retroviral sero-positive and had a CD4⁺ count of 39 cells/μl. She was then referred to the Retroviral Clinic where she is presently being managed for HIV/AIDS.

CASE 3

AJ, a 17yo student presented to our Clinic on account of inability to properly close his right eyelid, deviation of the mouth to the left side and slurred speech for 2 weeks. He also experienced overflow of tears. Patient said symptoms were noticed on waking up and had no previous history of any systemic illness (diabetes mellitus or hypertension) or trauma. There was no history of diarrhea or weight loss. At presentation, visual acuity in the RE was 6/18 and improved with pinhole to 6/9; VA in the LE was 6/5. There was left lagophthalmos, however extra-ocular motility was full range in both eyes. Other ocular examination findings were normal. A diagnosis of Bell's palsy was made and patient was commenced on systemic steroids 20mg daily; and advised to tape the right lids when sleeping. He re-presented at the Clinic two weeks later because he was experiencing daily transient visual loss that lasted between 15-30 minutes. About the same period, he noticed a papular rash on his mid forehead and upper lips. Retroviral screening was then requested for and proved to be sero-positive with a CD4⁺ count of 768 cells/μl. Patient is currently being managed for HIV/AIDS.

DISCUSSION

In a study involving 50 hospitalized patients with AIDS, signs of central eye movement limitation (56%) were the most common finding, followed by peripheral eye movement limitations, abnormalities of vision and abnormal spontaneous eye movements.⁶ This is similar to one of our cases who presented with mixed vertical and torsional nystagmus even though extra-ocular motility was normal. Other studies have also reported abnormal neuro-ophthalmological examination in most (60%) of their HIV positive patients - eye movement disorders were present in 51% of patients; others were visual field defects, optic neuropathy, papilledema, and ocular motor nerve palsies.⁹ In a study on the ocular complications of HIV/AIDS, Jads et al. reported that neuro-ophthalmic lesions were present in 8% of AIDS patients but did not however state the types of disorders seen.⁷ Bekele et al. reported cranial nerve palsies in only 2% of their patients. Their study however reported a significant relationship between ocular manifestation, sex

and age; and CD4+ count. CD4+ count <200 cells/μl and age >35 years were independent risk factors for ocular manifestations. Subclinical dysfunction in the visual pathways has been reported to be a common phenomenon in both HIV infected patients with and without neurological symptoms, but neurologically symptomatic patients seem to have more damage to their visual pathways.⁹

Similar to our first case (case 1), Cohen and Kardon¹¹ reported a case of idiopathic recurrent neuro-retinitis in a 29yo female based on a constellation of ocular findings including an RAPD, a prominent macular star and optic disc edema with normal laboratory investigations for syphilis, cat-scratch disease, complete blood count, Lyme titers, and angiotensin converting enzyme. In their case, the symptoms started with the right eye, underwent spontaneous resolution but recurred four years later involving the left eye. In our case, even though the recurrence and worsening of symptoms involved the same eye, it is likely that our patient had recurrent idiopathic neuro-retinitis because the clinical features resemble those reported by Cohen and Kardon.¹¹ The fact that all laboratory tests were within normal limits further points towards an idiopathic pathology.

Nystagmus is not a common ocular complication of HIV/AIDS, it is therefore not surprising that it initially presented a diagnostic dilemma because at presentation the patient appeared healthy and the nystagmus was of sudden onset. In this patient, despite a very low CD4+ count (CD4+=39 cells/μl), she was asymptomatic until the horizontal diplopia, and paraesthesia and dizziness that developed about 6 weeks after she first experienced the diplopia. Even though this patient later developed uveitis, this was not until about 6 weeks after the neuro-ophthalmic features. In a cross-sectional study of 40 HIV-positive patients in rural India, Gogri et al. found no significant association between neuro-ophthalmic lesions and CD4-count.¹² In their study, 10% of the patients had neuro-ophthalmic complications (papilledema, optic neuritis and 3rd cranial nerve palsy) with 75% having CD4+ counts >200 cells/μl and only one patient with papilledema had a CD4+ count of <100 cells/μl. This may explain why they found no significant association between CD4+ count and neuro-ophthalmic complications in their patients, as neuro-ophthalmic complications are known to be commoner with lower CD4+ counts.

CMV retinitis is known to be associated with low CD4+ count;¹³ this was however not the case with our patient, who despite a very low CD4+ count had normal fundal appearance. In a prospective cross-sectional study of 132 AIDS patients to determine the

prevalence of cytomegalovirus retinitis, Kuppermann et al.¹³ disclosed that in patients with CD4+ counts ≤ 50 cells/ μ l, about one third presented with CMV retinitis, whereas in those with CD4+ > 50 cells/ μ l, there was no CMV retinitis noted. They concluded that there was an increased risk of CMV retinitis with very low CD4+ counts. It is therefore surprising that our patient with a CD4+ count of 39 cells/ μ l did not manifest CMV retinitis – the reason for this is however not immediately apparent.

The third case was initially diagnosed as Bell's palsy but then when he presented later with transient visual loss and facial popular rash, he was further investigated and that was when he was found to be retroviral sero-positive.

CONCLUSION: These cases bring to the fore the importance of entertaining a high index of suspicion in managing unusual cases that present to the ophthalmologist; and in all cases to rule-out HIV/AIDS since it has its highest prevalence in sub-Saharan Africa. That way, early intervention can be instituted and this would ultimately reduce morbidity and mortality from the disease.

Consent Disclaimer:

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

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