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Case Report

Acute Retinal Necrosis Revealing HIV Infection in an 11 Year Old Girl

Nécrose rétinienne aigue révélatrice d'une infection à VIH chez une fille de 11 ans

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ABSTRACT

Acute retinal necrosis (ARN) may reveal immunosuppression from HIV infection. The authors report a case in an 11-year-old girl who was referred from a health centre for progressive decreasing vision in the left eye. Examination of this eye revealed a blind eye from complicated panuveitis. She tested positive for HIV. Anti-retroviral therapy was started after a paediatric consult. Two weeks later, she developed ARN in the right eye. Response to systemic antiviral therapy was favourable. Bilateral blindness was prevented due to prompt and adequate treatment. We recommend routine HIV testing in all patients with uveitis.

RÉSUMÉ

La nécrose rétinienne aigue peut révéler une infection à VIH. Nous rapportons le cas d'une patiente de 11 ans, référée d'un centre de santé pour une meilleure prise en charge d'une baisse progressive de l'acuité visuelle à l'œil gauche. L'examen de cet œil a montré qu'il était aveugle des suites d'une panuvéite. La sérologie HIV demandée est revenue positive. Par la suite, l'enfant a été vue par un pédiatre qui l'a mise sous un traitement antirétroviral. Deux semaines plus tard, elle a développé des plages de nécrose rétinienne à l'œil droit. L'évolution sous traitement antiviral par voie générale a été favorable. Ceci a permis d'éviter la cécité bilatérale chez cet enfant. Nous recommandons la recherche systématique du VIH dans les uvéites.

INTRODUCTION

Acute Retinal Necrosis (ARN) was described for the first time by Urayama *et al* in 1971 [1]. The Executive Committee of the American Uveitis Society refined the definition of ARN based on clinical characteristics and disease course to include: one or more foci of retinal necrosis with discrete borders located in the peripheral retina; rapid progression in the absence of antiviral therapy; circumferential spread; evidence of occlusive vasculopathy with arterial involvement; and a prominent inflammatory reaction in the vitreous and anterior chamber [2]. The disease is caused by an acute infection with a member of the herpes virus family. The presence of herpes simplex 1 and 2 was demonstrated by Culbertson *et al* after electronic microscopy and culture of the retina of an enucleated eye affected by ARN [3]. Cases of ARN from Varicella-Zoster virus (VZV) [4-6] and Cytomegalovirus (CMV) [7] have been reported. No case of ARN has to our knowledge been reported in Cameroon. We report a successfully managed case in which both the functional and vital prognosis were at stake.

CASE PRESENTATION

An 11-year-old girl was referred to our unit for rapidly decreased visual acuity in the left eye. She reported a two-week history of pain and redness in the left eye with rapidly progressive deterioration of vision. Past ocular history revealed that the left eye had intermittent redness in the 9 months preceding consultation. No medical care was sought.
Past systemic history was significant for chronic fungal infection of the fingernails. There was no recent history of illness including chickenpox or any herpetic skin lesion.

Ophthalmic examination revealed a visual acuity of 10/10 in the right eye and no light perception in the left eye. Examination of the right eye was normal. The left eye showed diffuse conjunctival injection, corneal oedema, non-granulomatous keratic precipitates, 2+ anterior chamber cells, unreactive dilated pupil, and a white cataract. The fundus was inaccessible. Intraocular pressure was 12 mmHg in the right eye and 5.3 mmHg in the left eye. Retinal detachment was seen on B ultrasound of the left eye. A diagnosis of panuveitis complicated by cataract and retinal detachment of the left eye was made. Dexamethasone eyedrops was given every two hours in the left eye.

The work-up included fasting blood sugar (FBS), complete blood count (CBC), erythrocyte sedimentation rate (ESR), CMV serology, HIV serology, CD4+ lymphocyte cell count and chest X-ray. VZV and HSV serologies were not done due to the unavailability of these tests in our milieu. The results were significant only for microcytic hypochromic anaemia; elevated ESR (99 mm at the first hour); positive CMV serology; positive HIV serology and CD4+ lymphocyte count of 144 x 106/l.

Anti-retroviral therapy (Abacavir, Lamivudine and Efavirenz) and clinical monitoring were started after a paediatric consult. Prophylactic therapy against opportunistic infections with Cotrimoxazole was also started. Two weeks later, she complained of floaters in the right eye. Examination of this eye revealed a visual acuity of 8/10. Slit lamp examination revealed 2+ anterior chamber and keratic precipitates. Dilated fundoscopy revealed moderate vitritis, peripheral retinal vasculitis with vascular sheathing; peripheral confluent retinal infiltrates with areas of retinal haemorrhage and the posterior pole appeared normal (figure 1).

Based on the clinical presentation and a positive CMV serology, we made a diagnosis of ARN. Vitreous tap was not done because of non-availability of polymerase chain reaction (PCR) of viral DNA. Liver functions, blood urea/creatinine were normal. Treatment was initiated as follows: peribulbar injection of dexamethasone; dexamethasone eye drops, given every hour; atropine eye drops 1%, given twice daily; and oral acyclovir 200 mg thrice daily.

Two days later, there was decreased vision (6/10); as well as circumferential and posterior spread of the retinal necrosis (figure 2). Intravenous acyclovir was started at 10 mg/Kg/8 hours. Forty-eight hours later, oral potassium supplement and high dose prednisolone (1 mg/kg/day) were started.

The response to treatment was marked by decreasing vitritis, regression of retinal necrosis with patchy retinal pigment epithelium atrophy on day 10 (figure 3). Intravenous acyclovir was switched to oral therapy. One month later, the vision improved to 10/10, steroid was slowly tapered off over three months. Prophylactic dose of oral acyclovir was maintained with regular monitoring of visual acuity, funduscopy, liver enzymes and kidney function. At one-year follow-up visit, there was no recurrence of ARN. Oral acyclovir was stopped and the patient was seen three months later with no recurrence. Monitoring continues regularly at 3 months interval and the patient and guardian were counselled to seek for immediate care in the case of any symptom.

DISCUSSION

Acute retinal necrosis (ARN) is an uncommon, but potentially blinding disease. Childhood ARN has not been previously reported in our setting. Only two cases of ARN in patients under the age of 18 years were seen in the Retina Service of Massachusetts Eye and Ear Infirmary over a ten-year period [8].

ARN is usually a disease of immunocompetent persons [9], but can also occur in immunocompromised individuals; as in this case. Immunocompromised patients are reported to have more severe and bilateral disease [10-12]. Batisse *et al* in a report on 26 cases of ARN in HIV-infected patients, reported bilateral disease in 59% [12]. ARN is a late event in the course of immunosuppression from HIV (CD4+ lymphocyte count < 100 x 10^6/l [11]. The CD4+ lymphocyte count of our patient was 144 x 10^6/l.

The diagnosis of ARN is made on a clinical basis [2]. Polymerase chain reaction (PCR) analysis for detection of viral DNA on ocular fluids is a reliable technique for the etiologic diagnosis. Negative HSV serology on ocular fluids does not exclude HSV-ARN [13]. We did not tap ocular fluid for serology in this monocular patient in whom signs were worsening and urgent treatment was required because of the absence of PCR analysis in our milieu.

Intravenous acyclovir is the mainstay of treatment. Newer agents like valacyclovir (oral) and forscanet (intravitreal or intravenous) can also be used [14, 15]. No prospective studies have investigated the optimal duration of intravenous treatment, timing of switching to oral therapy, or ideal total treatment duration. Patients with ARN are continued on prophylaxis with oral acyclovir after the retinal inflammation had subsided. Retinal detachment (traction and rhegmatogenous) is a common complication. This usually occurs within 2 months [8]. There is no consensus on the length of prophylaxis, which can range from months to a year [16]. Taking into consideration that our patient is monocular and risks bilateral blindness in case of recurrence, we maintained prophylaxis for one year and regularly examine the peripheral retina for any atrophic hole that can predispose to retinal detachment.

The role of the ophthalmologist in diagnosing and managing patients with HIV infection cannot be overemphasized, as ocular manifestations may be the presenting sign in an HIV-infected individual as reported in this case. Ebana *et al* reported that an ophthalmic manifestation was the inaugural manifestation of HIV in 31.6% of patients [17].

CONCLUSION

The diagnosis of ARN in this case was made after monocular blindness and involvement of the second eye. Prompt and adequate management in a multidisciplinary team approach led to a good outcome. We therefore recommend all health care givers to promptly refer any patient with visual symptoms to an ophthalmologist for detailed examination and proper management. HIV testing should be done routinely in all patients with uveitis.

DISCLOSURE

The authors declare that there is no conflict of interest regarding the publication of this paper.

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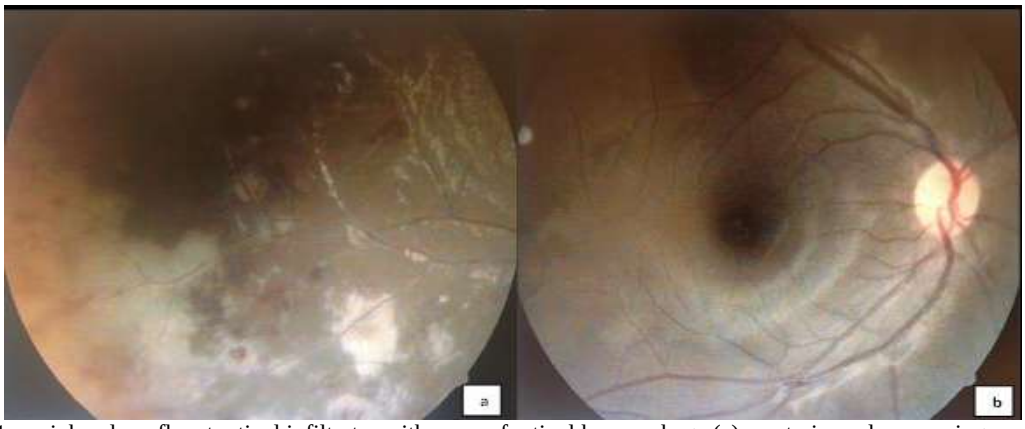

Figure 1: peripheral confluent retinal infiltrates with areas of retinal haemorrhage (a); posterior pole appearing normal (b)

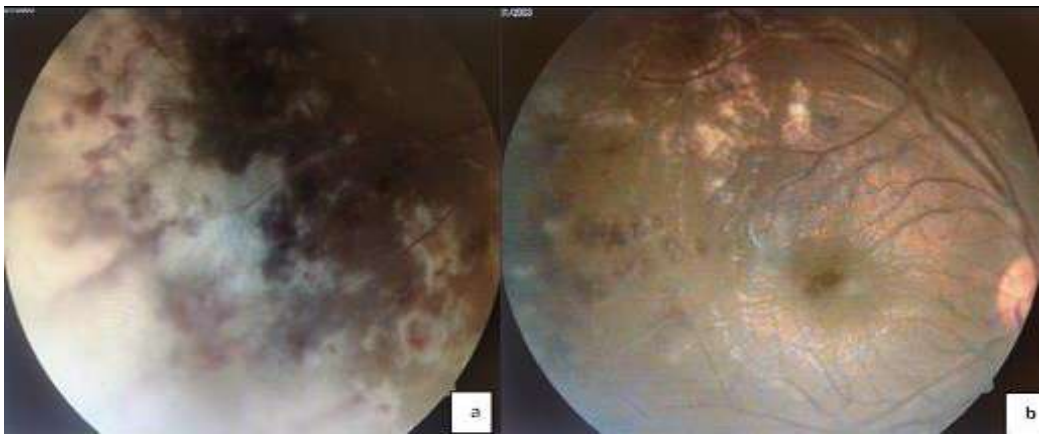

Figure 2: worsening retinal necrosis and haemorrhages (a) with spread of infiltrates and haemorrhages to the posterior pole (2).

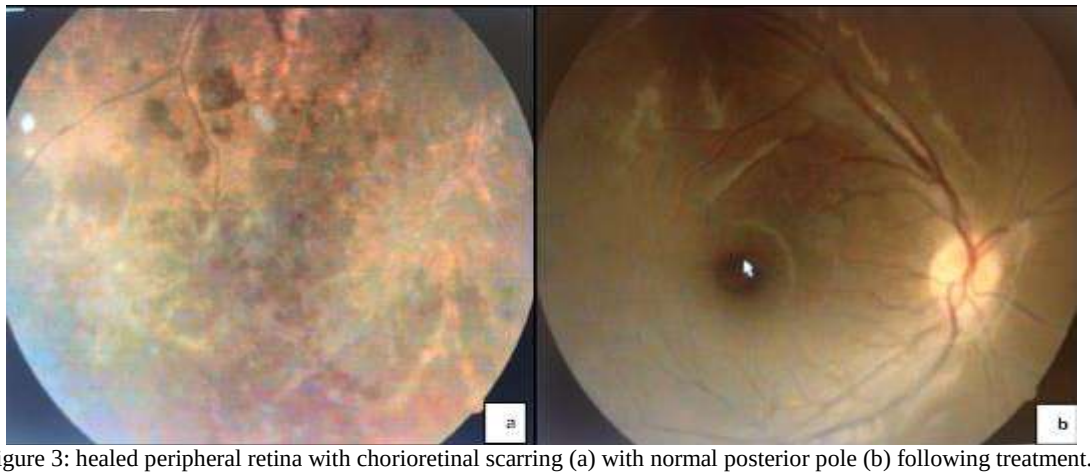

Figure 3: healed peripheral retina with chorioretinal scarring (a) with normal posterior pole (b) following treatment.