Case Report

Acute Retinal Necrosis Syndrome: A case report

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The acute retinal necrosis (ARN) syndrome represents a specific pattern of clinical presentation for certain herpes virus infections in the posterior segment of the eye. First described in the Japanese literature in 1971 and termed Kirisawa uveitis, it has till today remained a visually devastating condition in most of the cases. A female adult patient presented to us with classical picture of this syndrome and was managed with oral acyclovir and corticosteroid that responded well with this modality of treatment.

Introduction

Acute retinal necrosis (ARN) syndrome, first described in Japanese literature by Uramay et. al[1], is a visually devastating disease occurring in immunocompetent individuals which was later attributed to reactivation of dormant varicella zoster virus (VZV)[2] and herpes simplex virus, type I and type II[3]. This eventually leads to deterioration of visual acuity from classically described triad consisting of (1) an arteritis and phlebitis of the retinal and choroidal vasculature, (2) a confluent, necrotizing retinitis that preferentially affects the peripheral retina, and (3) a moderate to severe vitritis and the consequent late sequelae of retinal detachments[4-5] that occur in 75% of the cases and other complications. In US, 5.5% of uvietis for over a 10-year period is accounted for by ARN and 1.7% of uvietic cases in Switzerland[9]. No racial predilection exists. This condition appears to have a predilection for males but not certain to what extent. Occurrence of ARN has shown to have bimodal age distribution with herpes simple type II presenting at age of 20 years, and herpes simplex virus type 1 or varicella-zoster virus group with median age of involvement at 47 years and 57 respectively.

Untreated bilateral retinitis occurs in 80% of cases and fellow eye involvement occurs in 90% of these cases in first 3 months even though interval of 26 years has been reported.

Treatment entails use of intravenous acyclovir[6] expecially to hasten resolution of the infection and prevent second eye involvement[4], systemic and topical corticosteroid, antithrombotic agents, prophylactic photocoagulation of the atrophic holes, and vitrectomy with acyclovir drip with gas or oil tamponade of the detachment, and optic nerve decompression in case of its involvement [7-8].

Case Report

Fifty-one years old female patient presented to outpatient of BPKLCOS, Tribhuvan University Teaching Hospital on 4th August 2003 with history of redness in the left eye since 20th of July 2003, followed a week later with blurring of vision, visualization of fine floaters and a dull pain in and the around eye. There was no other significant ocular problems that occurred either preceding the onset or during the initial course of this illness. Systemically, she did not have any fever, eruptions or rashes, cough or any associated diseases. She had been wearing presbyopic glass for the last 3 years.

Figure 1: Fifty-one years old Tamang woman patient.

Examination revealed that she was a healthy woman with no evidence of suffering from any concurrent systemic diseases.

On ophthalmic evaluation, her vision in the right eye was 6/6 with [+0.75/+0.50x180°] and N/6 with +2.00 diopters sphere but in left eye 1/60 without any improvement. Extraocular movements were full. Right eye was essentially normal in both anterior and posterior segments evaluation.

The anterior segment in the left eye revealed minimal circumciliary congestion with large to moderate size KPs clustered in the inferior and center of the endothelium, flare of 2 +, cells of 3 +, iris was of normal pattern and color, pupil was round with regular margin and the crystalline lens had iris pigment deposits in the periphery especially in the superionasal part. Vitreous revealed grade 2 haze with 4+ cells and opacities. Optic disc was elevated with blurred margins, severely edematous and hyperemic. All the visible branches of retinal artery had features of severe arteritis. Phlebitis of smaller branches of retinal vein with tortuosities were noted. Peripheral retina had yellowish pale confluent patchy areas of retinal necrosis extending from 3 to 8 clock hours, more marked in inferiotemporal areas with sharp demarcation from normal retina.

Figure 2: Moderate to large size KPs with flare.

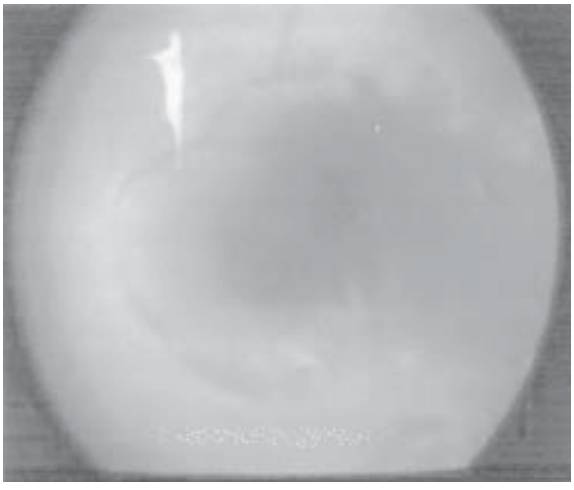

Figure 3: Fundus picture of the affected left eye revealing severely edematous disc, obliterative retinal arteritis and phlebitis, macular edema and generalized retinal edema.

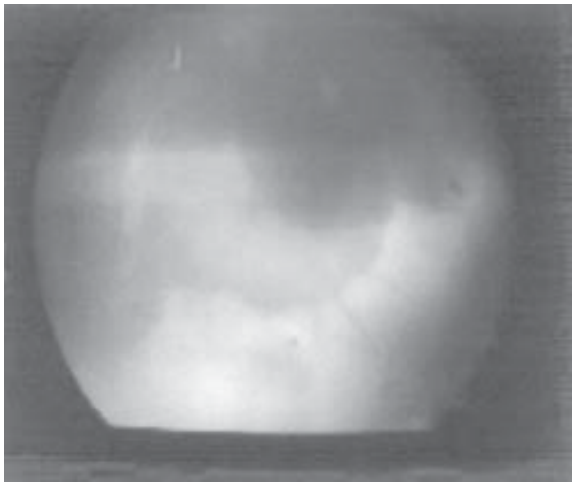

Figure 4: Inferiotemporal area with active peripheral retinal necrosis that has sharp demarcation with the uninvolved retina.

Intraocular pressure was 9 mm Hg in the right eye and 8 mm Hg in the left.

With the clinical diagnosis of left eye ***Acute Retinal Necrosis Syndrome*** she was immediately admitted and started on oral acyclovir 800 mg five times a day. Topical Predacetate 1% one hourly and atropine drop 1% in the left eye three times were also commenced and investigations conducted.

Ultrasound revealed scattered multiple fine echo dense shadows in vitreous, thickened optic nerve and attached retina. The FFA gave a picture of delayed filling from the choroidal phase and prolonged arteriolar phase. There were areas of patchy filling defects in periphery.

The hematological tests showed Hb of 13.3 gm%, white blood cells count of 12,900/ cu mm, ESR of 10 mm 1st hour, differential count of 83% Neutrophils, 15% of lymphocytes and 1% each of Monocytes and Basophils. The random blood sugar was of 4.6 mmol/L and renal and liver function tests were of normal values. TORCH IgM ELISA tests were negative and both HIV and VDRL tests were non-reactive. In the TORCH IgG antibody ELISA tests, Toxoplama gondii was 1000.00 IU/ml (N=10.0 IU/ml), rubella with 20.0 IU/ml (N=10.0 IU/ml), CMV of 2.8 IU/ml (N=0.25 IU/ml) and positive HSV I but negative for HSV II. Urine was normal in routine and microscopic examination and stool revealed presence of hookworm ova. Chest X-ray PA was essentially normal.

On 8th August 2003, oral steroid was started at 1 mg/kg body weight along with daily one tablet of 325 mg aspirin in conjunction with ranitidine 150 mg per oral twice daily.

On 10th of August 2003, she had regained vision to 5/60 improving to 6/36 with pinhole. Anterior segment showed

reduction in KPs, cells and flare and similar observation in vitreous. No significant change was noticed in the fundus picture.

Findings of left eye on 25th August 2003 revealed, her visual acuity of 6/60 improving to 6/36 with pin hole, minimal fine KPs some of which are pigmented and crenated, occasional cells in AC with flare 1+, no alterations in iris or lens, vitreous showed cells 2+ with haze of 2+ and opacities 3+. Fundus still revealed disc swelling with sclerosed branches of retinal artery. A small patch of ischemia noticed inferior to macula threatening the central vision. The lesions in the peripheral retina showed evidence of gradual regression.

After more than 6 weeks, on 11th September 2003, the patient was discharged. Her visual acuity had returned to 6/36 improving to 6/24 with pinhole in the left eye. No fellow eye involvement noted. The anterior segment involvement had abated almost completely and so do the reactions in the vitreous. Still, the disc had blurry elevated margins with pale appearance. Outline of occluded arteries were noted more distinctly but surprisingly veins appeared to have sustained less damage. Macular edema was still present but the ischemic patch appeared less prominent. The involved retinal periphery was being replaced with patchy areas of atrophic scars. No obvious areas of detachment or holes visible at present.

Her latest follow up was on 24th November 2003. Visual acuity in right eye was 6/6 with +0.75 DS and left 6/18 with +1.5 DS. Cornea was clear with few scatttered central pigment deposits, quiet anterior chamber, oval pupil with posterior synechiae but reacting to light, clear lens, vitreous haze had disappeared. Fundus showed mild disc pallor with inferiotemporal sclerotic vessels, pigmentary changes in macula. IOP was 13 mm Hg in right and 9 mm Hg in left. Peripheral retina had atrophic changes. Right eye was essentially normal.

Discussion

This patient presented in stage 1b with confluent areas of peripheral retinitis, papillitis, and macular edema going on to stage 2 with vitreous opacification or organization. Stage 3 was noted after 4 weeks of onset.

Even though, intravenous acyclovir is the mainstay of therapy given at 1500mg/m2 every 8 hours for 7-10 days followed by oral acyclovir (2-4gm daily) usually for 4-6 weeks [4], she received a full 6 weeks course of oral therapy without any untoward complications or adverse reaction. Clinical response was found to be effective from the improvement noted in this patient. The role of steroid was controversial but in this patient it was given due to massive papillitis.

Therefore, administration of adequate oral antiviral therapy gives good response as demonstrated by this case. Immediate institution of therapy along with regular monitoring of the progress and timely detection of the involvement in other eye should be done. Prophylactic or any surgical interventions were not found to be necessary in this case.

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