Acute Retinal Necrosis Following Herpes Simplex Encephalitis

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Acute retinal necrosis is a severe ocular inflammatory syndrome associated with a very poor visual outcome. Though it is known to occur occasionally in association with or shortly after herpetic encephalitis, 1,2,3 ARN occurring years after encephalitis has been rarely reported. 4,5 Here we present a case of unilateral ARN occurring 5 years after herpes simplex virus encephalitis.

Case Report

A 14 year old male presented with redness and watering of his right eye of a week’s duration. He had been on local steroids and mydriatics for 3 days. He gave a history of herpes encephalitis, with post viral demyelination, cystitis and septicaemia 5 years back. At that time he had presented with headache, fever, vomiting, slurring of speech, drooling of saliva and inability to feed. Though blood and cerebrospinal fluid titres of antibodies to herpes simplex virus were negative, he was diagnosed to have herpes encephalitis on the basis of CT scan and MRI findings which showed diffuse brain oedema. He responded well to antiviral therapy and had been asymptomatic since then.

On presentation, the best corrected visual acuity was 6/18, N8 in the right eye and 6/6, N6 in the left eye. Anterior segment showed evidence of anterior uveitis in the right eye with keratic precipitates, 2+-cells and 1 + flare. Left eye was normal. Dilated fundus showed vitritis with large whitish yellow confluent retinal

Fig 1. (a) showing vitritis and normal posterior pole right eye (b) showing confluent retinal infiltrates and vasculitis in the periphery of the right eye
infiltrates and vasculitis in the retinal periphery in the right eye (Fig 1 a & b). The retina in the left eye was normal.

A diagnosis of acute retinal necrosis was made and he was started on T. Acyclovir 800 mg 5 times and T. Wysolone 60 mg daily along with topical steroids and mydriatics. Investigations performed were IgG and IgM antibody titres for Herpes Simplex virus, Cytomegalovirus, Varicella Zoster and Human Immunodeficiency virus. He was advised review every 2 days. Investigation results revealed positive antibody titres for IgG and IgM for HSV and IgG for CMV. The lesions were found to be remaining status quo without any increase in size (Fig 2 a & b)

He was kept on regular follow up with Acyclovir and steroids in tapering dose once the lesion started resolving. One month later best corrected visual acuity was 6/9, N12 in the right eye and 6/6, N6 in the left eye. Fundus showed resolved ARN with healing vasculitis (Fig 3 a & b).

He was advised to stop T. Acyclovir and Wysolone and to continue topical steroids and topical flurbiprofen eyedrops. However one month later he once again presented with recurrence of panuveitis and vision drop to 6/18 and had to be restarted on steroids and Acyclovir. Despite treatment, the vitritis increased though retinal lesions did not recur (Fig. 4 a & b). An epiretinal membrane had formed at the macula. A therapeutic pars plana vitrectomy under general anaesthesia was done along with peeling of epiretinal membrane (Fig. 5) and prophylactic barrage laser of the peripheral retina. The vitreous aspirate was sent to

Fig. 2 a & b showing lesions remaining status quo without increase in size or number

Fig 3 a & b showing resolved ARN with healing vasculitis after one month Right eye
detect IgG and IgM antibodies for Herpes Simplex virus, Cytomegalo virus, Varicella Zoster virus. IgG antibody for HSV1 was found to be positive. Postoperatively the eye was quiet with clear fundus view and normal retina. On review after 1 month the eye remains quiet with no recurrence and a best corrected visual acuity of 6/9, N8. (Fig. 6 a & b)

**Discussion**

ARN (Acute Retinal Necrosis) was first described by Urayama et al in 1971 in 6 patients with severe intraocular inflammations, retinal vascular sheathing and large white confluent retinal infiltrates which progressed to rhegmatogenous retinal detachment.
Acute retinal necrosis syndrome or ARN is characterised by initial onset of episcleritis or scleritis, periorbital pain, and anterior uveitis. This is followed by decreased vision resulting from vitreous opacification, necrotizing retinitis and in some cases optic neuritis or neuropathy. The retinitis appears as deep multifocal, yellow-white patches in the peripheral fundus which become concentrically confluent and spread towards the posterior pole; the macula is frequently spared. An active vasculitis is present with perivasculares hemorrhages, sheathing and terminal obliteration of arterioles by thrombi. This phase of active retinitis usually lasts for 4 to 6 weeks during which time an exudative retinal detachment may occur.

With resolution, pigmentation of the lesions begin at their posterior margins, leaving a scalloped appearance. Retinal breaks may develop at the junction of normal and necrotic retina. A rhegmatogenous retinal detachment or tractional retinal detachment due to organisation of vitreous inflammation and proliferative vitreoretinopathy may occur.

The contralateral eye is involved in about 36% cases usually within 6 weeks of onset in the other eye though it may be affected after as long as 34 years. ARN commonly occurs in otherwise healthy patients of either sex and of any age. ARN can also occur in immunocompromised patients. Immunocompromised patients have been described to have skin manifestations of zoster and CNS involvement.

Considerable evidence points to one or more members of the herpes virus family in the etiology of ARN syndrome. Varicella –zoster virus, cytomegalovirus, herpes simplex have all been implicated. Some studies suggest that Varicella –zoster virus and Herpes simplex virus type-I cause ARN in patients older than 25 years, whereas herpes simplex virus type-2 causes acute retinal necrosis in patients < 25 years.

Retinal necrosis in ARN probably is the result of multiple factors like direct lytic viral infection of retina, immune complex disease mediating an obliterator vasculitis and vitreous inflammation and traction. This case validates the theory of brain to eye transmission of the virus through the optic nerve. Thus, following encephalitis retinal neurons may function as a reservoir for HSV that can be reactivated to cause ARN several years later. The occurrence of ARN would support the etiological suspicion of the previous encephalitis.

Treatment is with acyclovir which has good activity against HSV and VZV. It can hasten resolution and prevent contralateral spread. Dosage is 1V 500 mg/m² every 8th hr for 10-14 days or orally 800 mg x 5 times for 6 weeks. Valacyclovir orally 1gm tds or famicyclovir 500 gm tds have the same efficacy and lesser side effects. In case of severe progression despite acyclovir, ganciclovir or foscarinet can be given. Systemic corticosteroid therapy in a dose of 0.5-1 mg/kg is given after 24-48 hours of IV acyclovir.

Aspirin is given in extensive arteritis and retinal vascular occlusion. Other modalities of treatment are intravitreal injection of antiviral, barrage laser photocoagulation, prophylactic scleral buckling with vitrectomy, endolaser photocoagulation and silicone oil tamponade in eyes at risk etc.

ARN resolves spontaneously in 2-3 months with or without therapy. However retinal detachment with large breaks with rapid progression of the disease can occur in untreated cases.

In this case, a previous history of herpetic encephalitis was useful in making the diagnosis of acute retinal necrosis and initiating treatment at the earliest. Thus though the disease is associated with poor visual prognosis, prompt treatment could prevent complications from occurring as in this case.

**Conclusion**

Pediatricians and physicians should be aware of this entity and be alert to recurrences that may be delayed by years. Similarly previous history of central nervous system infection can help to clinch the diagnosis of ARN. Earliest diagnosis and treatment is important to prevent complications of ARN.

**References**


HUMOUR IN OPHTHALMOLOGY

In the lighter vein

Dr. Varma MS

Dr. Bhikhalal S. Patel’s name may not be in any Ophthalmic Hall of Fame. But I am sure; it will be engraved in the minds and hearts of generations of alumni of B.J. Medical College, Ahmedabad.

Affectionately called “Bhikhu-bhai” by all and sundry, he was the honorary head of the second unit of Ophthalmology of that 128-year old college during seventies and eighties. He was not a great clinician or surgeon; nor was he a great teacher. In fact, he never taught us any Ophthalmology; he taught us about Life. And he taught not by pedagogy; but by his own life. He was the master of patient management. The way he built rapport with; consoled and comforted; cajoled and convinced; humoured and honoured patient after patient was a treat to watch. I once had the fortune to accompany him on a house visit; or so I thought.

One day as I was hurrying back to the OPD after a mid morning coffee break, he asked me to go with him to a nearby house to see an old patient of his. Since only he had an air-conditioned car in the department in 1980, I was quick to grab the chance. He drove to the nearby Asarva Housing Colony and stopped in front of a ‘two room – kitchen’ apartment. There were about fifteen people in the house. Their reaction was astounding. It was as if the Lord God himself had stepped into the house. And as God’s assistant, a little reflected glory fell on yours truly too. When the head of the house assured him that everything was ready, Dr. Patel asked me to fetch ‘that thing’ from his car. ‘That thing’ proved to be a stainless steel basin with five or six surgical instruments and a big ball of cotton in it. While we were served too sweet tea, the basin was filled with some water and set to boil on the stove. Then it struck me! He was going to do a cataract operation!

While I held a feeble torch, and all the family members crowding around, he did an ICCE/PI (knife section, no sutures!) in about fifty seconds flat. As soon as the cataract was out (capsule forceps) he gave it to the nearest relative saying “Lo! Mothiaam” (Here! The Cataract). It was reverently passed round. Then squeezing out the ball of cotton, he put it on as pad and bound up the eye. I would be omitting some truth if I do not add that while going out one of the relatives slipped an envelope (Rs.20/-) in my pocket. Assistant’s fees! Whenever the topic of deteriorating doctor-patient relations come up, I am reminded of my great teacher who inspired such blind faith in his patients.