

Case Report

Kyrieleis plaques associated with Herpes Simplex Virus type 1 acute retinal necrosis



Neha Goel*; Amrita Sawhney

Abstract

We report the case of a 55-year-old immunocompetent male who presented with features typical of acute retinal necrosis (ARN). Polymerase chain reaction of the aqueous tap was positive for Herpes Simplex Virus (HSV) – 1. Following therapy with intravenous Acyclovir, followed by oral Acyclovir and steroids, there was marked improvement in the visual acuity and clinical picture. At one week after initiation of treatment, Kyrieleis plaques were observed in the retinal arteries. They became more prominent despite resolution of the vitritis, retinal necrosis and vasculitis and persisted till six weeks of follow-up, when fluorescein angiography was performed. The appearance of this segmental retinal periarteritis also known as Kyrieleis plaques has not been described in ARN due to HSV-1 earlier.

Keywords: Kyrieleis plaque, Acute retinal necrosis, Segmental periarteritis, Herpes Simplex Virus type 1

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Introduction

Kyrieleis plaques are a rarely encountered clinical entity in which whitish segmented deposits are seen scattered along retinal arterial branches in a beaded pattern.¹ Also known as segmental periarteritis,² this feature has primarily been described in association with toxoplasmosis,³ tuberculosis,¹ syphilis⁴ and Mediterranean spotted fever.⁵ We present the occurrence and course of Kyrieleis plaques in acute retinal necrosis (ARN) due to Herpes Simplex Virus (HSV) – 1.

Case report

A 55-year-old immunocompetent male presented with decreased vision and floaters in his left eye since 10 days. There was no significant past medical history. Best corrected visual acuity (BCVA) was 20/20 in the right eye and 20/200 in the left eye. Slit lamp examination demonstrated granuloma-

tous anterior uveitis with 3+ cells in the anterior chamber in the affected eye. A dilated fundus examination of this eye revealed 2+ vitreous cells and patches of retinitis in the mid and far retinal periphery with neighboring vasculitis (Fig. 1). The right eye examination was unremarkable.

Polymerase chain reaction of the aqueous tap was positive for HSV-1 and negative for HSV-2, Varicella Zoster Virus (VZV) and Cytomegalovirus (CMV). A diagnosis of unilateral ARN was made. The patient was started on intravenous Acyclovir 500 mg three times a day for 7 days followed by oral Acyclovir 800 mg five times a day with oral Prednisone 60 mg daily. Topical steroids and cycloplegics were also administered in the left eye.

The patient demonstrated improvement within a week of initiation of therapy. At this stage, Kyrieleis plaques were observed along two inferior retinal arteries (Fig. 2a and d). At two weeks follow-up, BCVA improved to 20/60 with resolution of the anterior uveitis and decrease in the vitritis and

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ICARE Eye Hospital and Postgraduate Institute, Noida, U.P., India

* Corresponding author at: 57, Mayur Vihar Phase 1 Extension, New Delhi 110091, India. Tel.: +91 9811179191; fax: +91 11 23230033. e-mail address: nehadoc@hotmail.com (N. Goel).



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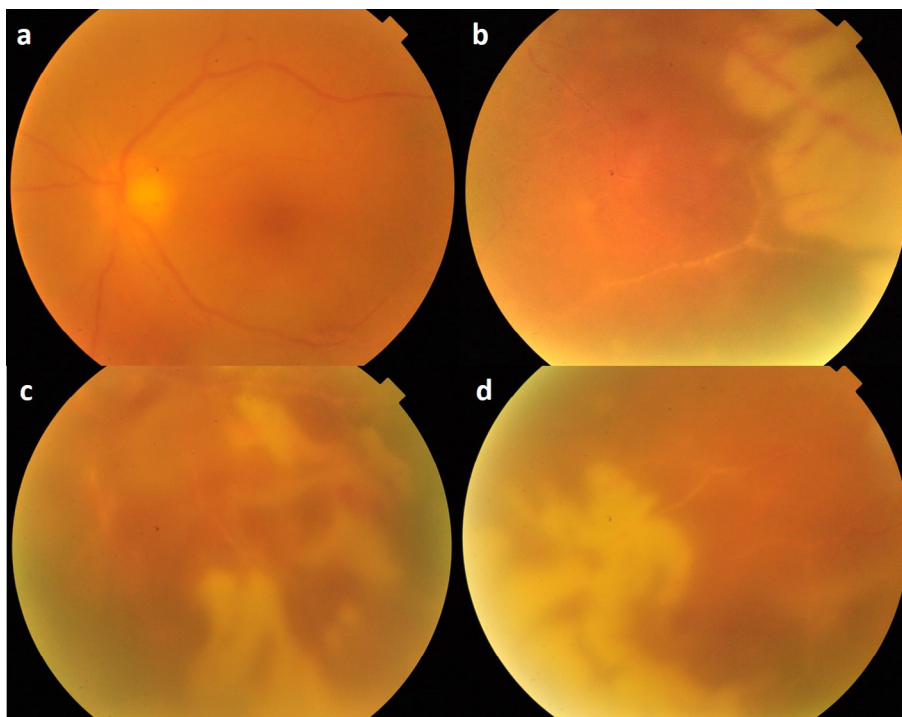


Figure 1. Colour fundus photograph of the left eye at presentation showing vitritis (a), peripheral areas of retinal necrosis with neighboring vasculitis in the temporal (b), inferior (c) and nasal quadrants (d).

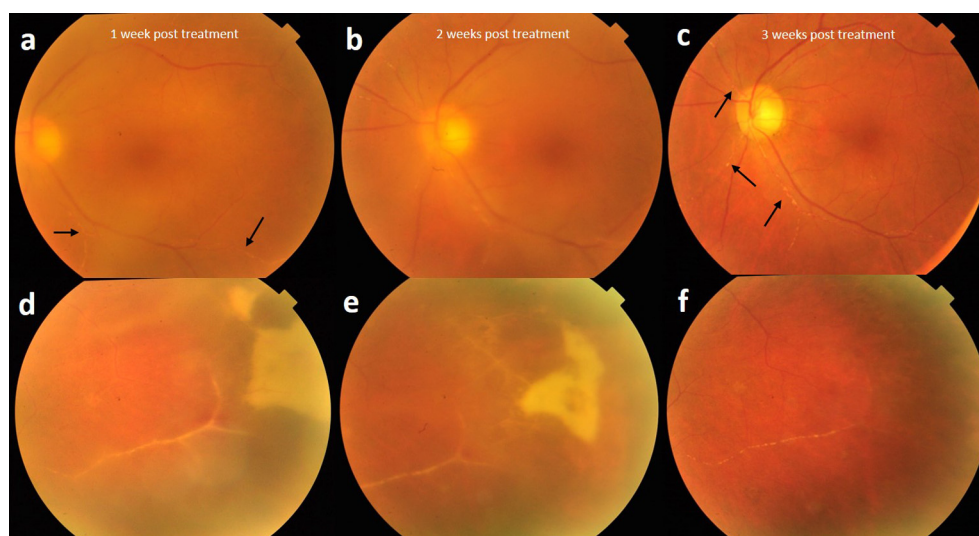


Figure 2. Weekly fundus photographs of the left eye following initiation of therapy. (a) After a week of intravenous Acyclovir, vitritis decreased and Kyrieleis plaques were visible in the inferior retinal arteries (arrows). (d) The borders of the retinal necrotic lesions became more well defined. The patient was shifted to oral Acyclovir and steroids. (b) A week later, there was further decrease in the vitritis and the retinal necrosis (e). Treatment was continued and at three weeks, there was resolution of the vitritis (c) and peripheral retinal necrosis (f). Kyrieleis plaques were seen in nasal retinal arteries as well, as yellowish plaques that did not extend beyond the vessel walls (arrows). There was no involvement of the retinal veins.

peripheral retinal necrosis (Fig. 2b and e). Three weeks later, BCVA was 20/30, vitritis had resolved and the retinitis was no longer active. At this stage, Kyrieleis plaques were more numerous and prominent, also observed along nasal retinal arteries (Fig. 2c and f). These persisted till six weeks follow-up (Fig. 3a), when fluorescein angiography was performed. There was no delay in arterial filling, no leakage from the retinal arterioles and the plaques themselves did not fluoresce (Fig. 3b and c). Staining was present in the areas of resolved

retinitis (Fig. 3d). Steroids were tapered and oral Acyclovir discontinued after two weeks.

Discussion

ARN is an uncommon intraocular inflammatory syndrome that typically affects immunocompetent individuals of all age groups. Clinically, it is characterized by anterior uveitis, dense vitritis, progressive retinal necrosis that begins in the

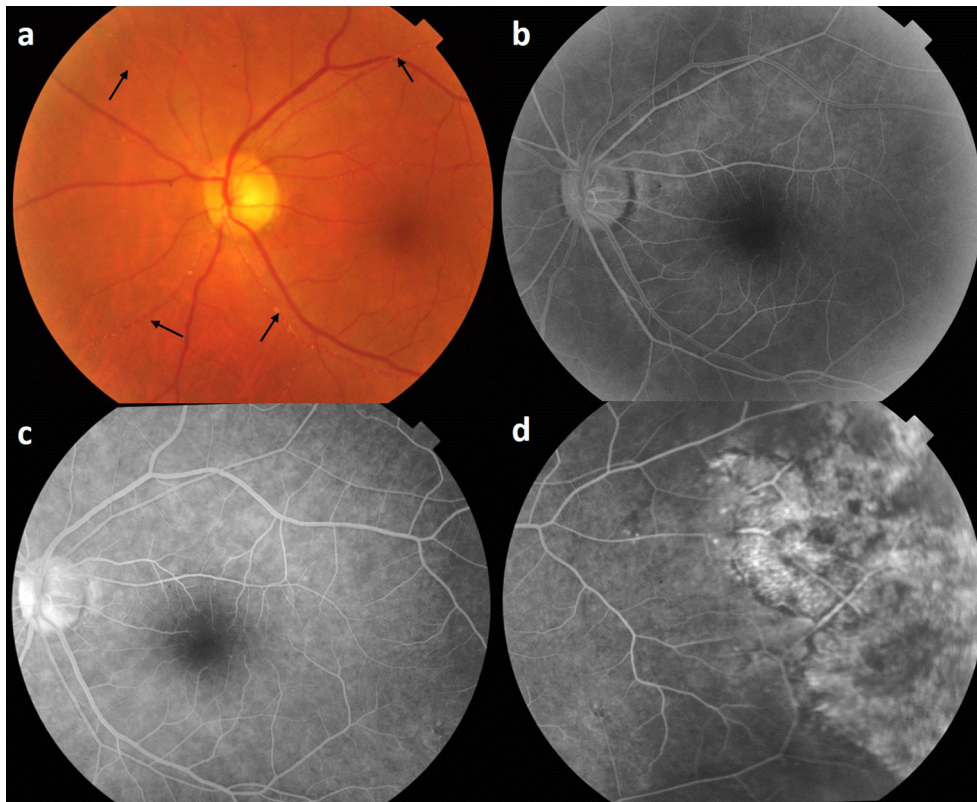


Figure 3. (a) At six weeks following presentation, fundus photograph of the left eye showing Kyrieleis plaques in the retinal arteries (arrows). Fundus fluorescein angiography did not demonstrate any leakage or staining of the retinal arteries in the areas of the plaques (b and c). The peripheral healed necrotic retinal areas showed staining (d).

periphery and occlusive vasculitis. The diagnostic criteria for ARN were established by the American Uveitis Society in 1994.⁶ ARN usually presents unilaterally and carries a poor prognosis. Visual loss occurs due to retinal detachment, optic neuropathy and ischemic vasculopathy involving the macula.⁷

The etiological agents of ARN include DNA viruses such as VZV, HSV and less commonly CMV. While HSV-2 is commoner in younger patients, HSV-1 or VZV is more prevalent in older patients.⁷ These can be identified by PCR-based assays of ocular fluids. However, ARN is principally a clinical diagnosis and institution of therapy should not be delayed while awaiting laboratory confirmation. Prompt diagnosis and treatment is essential to salvage vision in this frequently blinding condition and preventing involvement of the fellow eye. Treatment involves extended use of antiviral agents. Systemic steroids help to limit damage caused by the severe inflammation associated with ARN.⁷

Kyrieleis plaques were initially described in an eye with tuberculous uveitis by Kyrieleis in 1933.¹ The term "segmental periarteritis" was used later to describe these whitish segmented deposits found within retinal arteries.² Kyrieleis plaques have also been associated with infections of the retina due to toxoplasma, syphilis and *Rickettsia conorii*.³⁻⁵ Their occurrence in ARN is rare. There is a single case report of Kyrieleis plaques in ARN due to HSV-2,⁸ and two reports in ARN due to VZV.^{9,10} To the best of our knowledge, this is the first report of Kyrieleis plaques in ARN due to HSV-1.

Kyrieleis plaques can be differentiated from vascular sheathing and frosted branch angiitis. While the former affects retinal arteries exclusively, the latter can involve both

retinal arteries and veins. Also, the plaques do not leak fluorescein as opposed to frosted branch angiitis.¹¹

Since there is no pathological study of Kyrieleis plaques, their nature is still unknown. In our patient, these plaques appeared after one week of therapy, when the vitritis and retinitis were resolved and increased thereafter. A similar course has been observed in the previous cases.^{9,10} This suggests that they may represent an immunological response to an infectious agent resulting in the deposition of inflammatory debris within or adjacent to the vessel wall.¹² However, persistence of the plaques despite resolution of the infection and treatment with steroids contradicts this hypothesis.³

In summary, this case represents a typical ARN syndrome due to HSV-1 infection that demonstrated Kyrieleis plaques following treatment. Occurrence of Kyrieleis plaques may be more common than the literature suggests, and they may be underreported. Our case adds another cause to the etiology of these plaques.

Conflict of interest

The authors declared that there is no conflict of interest.

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