

Serpiginous choroiditis and acute retinal necrosis occurring in the same patient

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We describe a rare association of serpiginous choroiditis with necrotizing retinitis having clinical features of acute retinal necrosis (ARN). A 23-year-old male developed ARN in the fellow eye while he was on tapering doses of immunosuppressive medications for unilateral serpiginous choroiditis. The association may represent a common viral etiology of the two diseases or may be due to the development of ARN due to general state of iatrogenic immunosuppression. This report also highlights the importance of a detailed evaluation of both the eyes on regular follow-up visits in the patients receiving iatrogenic immunosuppression.

Key words: Acute retinal necrosis, immunosuppression, serpiginous choroiditis

Serpiginous choroiditis is rare inflammation of the retinal pigment epithelium and choroid; the macular form being well-recognized.^[1] The etiology remains unknown despite various studies implicating infectious agents including viral etiology in the pathogenesis.^[2,3]

Acute retinal necrosis (ARN) has established viral etiology and is characterized by presence of peripheral retinal necrosis with retinal arteritis and prominent vitritis in immunocompetent patients.^[4] However, the disease has been reported to occur in immunosuppressed patients, including iatrogenic immunosuppression.^[5,6] The diagnosis of ARN is usually based on clinical findings.^[4]

We report a patient with unilateral serpiginous choroiditis who developed necrotizing retinitis with clinical features of acute retinal necrosis in the fellow eye.

Case Report

A 23-year-old male presented with the complaint of decreased vision in the right eye since 3 months. He had been treated

elsewhere for right eye choroiditis with intravenous methyl prednisone acetate (IVMP) 1 g/day for 3 days followed by oral steroids and Tab Azathioprine. At presentation, he was taking Tab Prednisone 50 mg/day and Tab Azathioprine 125 mg/day. Best corrected visual acuities (BCVA) were 20/200 and 20/20 in the right and left eyes, respectively. Left eye was essentially normal on examination. Right eye revealed quiet anterior chamber and quiet vitreous cavity with a yellowish area of active choroiditis with geographic borders involving the macula in serpiginous pattern. Fundus fluorescein angiography confirmed the findings. He was diagnosed as a case of active macular serpiginous choroiditis. After obtaining physician's clearance, we prescribed Tab Prednisone 60 mg/day (1 mg/kg body weight), Tab Azathioprine 50 mg thrice a day, Tab Cyclosporine 150 mg twice a day, with antacid and calcium supplements. QuantiFERON-TB Gold test was found to be negative. On regular follow-up visits, the choroiditis lesions were noted to heal over a period of 3 months, and treatment was gradually tapered.

After 3 months, patient complained of redness, pain and blurring of vision in the left eye. At this point of time, he was on Tab Prednisone 7.5 mg/day, Tab Azathioprine 50 mg/day and Tab Cyclosporine 200 mg/day. BCVA were 20/400 and 20/20 partial in the right and left eyes, respectively. Right eye examination revealed a quiet eye with healed lesions of macular serpiginous choroiditis [Fig. 1]. Left eye had 2+ anterior chamber cells, severe vitritis, and an area of retinal whitening suggestive of necrotizing retinitis in the inferonasal periphery. A clinical diagnosis of acute retinal necrosis was made. Tab Cyclosporine and Tab Azathioprine were stopped, Tab Prednisone was hiked to 60 mg/day, and intravenous Aciclovir 500 mg thrice a day was started under monitoring. Serum ELISA for Human immunodeficiency virus (HIV) was negative. On regular examinations, vitreous haze decreased markedly and the lesion showed signs of healing [Fig. 2]. After 1 week, intravenous Aciclovir was stopped and the patient was prescribed Tab Valaciclovir 1 g thrice a day, and Tab Prednisone to be tapered at 10 mg/week. As on last visit after 2 months, anterior chamber was quiet and retinitis had resolved significantly. The patient is under regular follow-up.

Discussion

We report a patient who developed ARN in the fellow eye while he was on tapering doses of immunosuppressive medications for unilateral serpiginous choroiditis. To the author's knowledge, the association of serpiginous choroiditis with ARN has not been reported earlier. This association can be explained by two possibilities; it may represent a common viral etiology of the two diseases or development of ARN due to general state of iatrogenic immunosuppression.

The viral etiology for serpiginous choroiditis is not yet firmly established.^[2,3] Gass *et al.* reported a case of serpiginous choroiditis following herpes zoster ophthalmicus and suggested a possible viral etiology in some patients with serpiginous choroiditis.^[7] Priya *et al.* suggested that herpes viruses might have a role in the pathogenesis of the disease in a subset of patients with serpiginous choroiditis.^[2] Erkkilä *et al.* reported increased levels of antiviral (herpes simplex) antibodies in serum of two patients with serpiginous choroiditis.^[3] Ampigenous choroiditis, a variant of serpiginous

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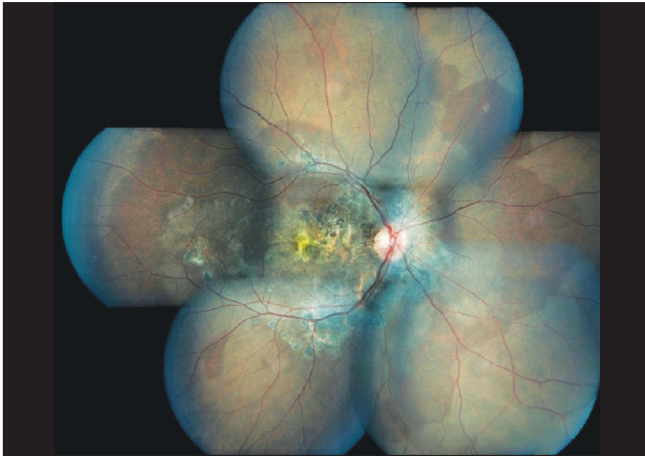


Figure 1: Color fundus photograph of the right eye showing the healed lesions of macular serpiginous choroiditis

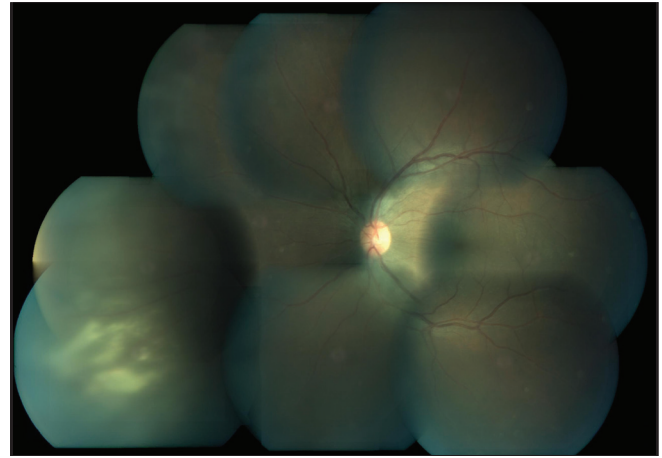


Figure 2: Color fundus photograph of the left eye showing presence of vitreous haze with white lesion of necrotizing retinitis (acute retinal necrosis) in the inferonasal periphery

choroiditis, has been reported to occur following quadrivalent human papilloma virus vaccine.^[8] The treatment of serpiginous choroiditis with systemic antivirals has been recommended and practiced by some authors.^[9,10] In our case, the limitation is the lack of serological diagnosis to prove a common herpetic origin of the serpiginous choroiditis in the right eye and necrotizing retinitis in the left eye. However, most immunologic explanations of serpiginous choroiditis deal with hypersensitivity reactions and virus would not need to be present in the eye in order to explain the disease. Likewise, though anterior chamber biopsy with polymerase chain reaction (PCR) testing for herpes virus would have strongly confirmed the diagnosis of ARN in the left eye, ARN is a clinical diagnosis and has an established viral etiology.^[4]

The other possibility can be the development of ARN due to iatrogenic immunosuppression. ARN has been reported to occur in immunosuppressed patients, including iatrogenic immunosuppression^[5] and following intravitreal steroid injections.^[6] Though our patient was not found to have an HIV infection, he had received a course of IVMP elsewhere and was on immunosuppressive medications for almost 9 months when he developed ARN.

The temporal relationship between serpiginous choroiditis and ARN, reported in the present case, is intriguing. Our case report also highlights the importance of a detailed evaluation of both the eyes on regular follow-up visits in the patients receiving iatrogenic immunosuppression.

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