CHICKEN POX ASSOCIATED BILATERAL RETINAL VASCULITIS IN AN IMMUNOCOMPETENT YOUNG MALE; AN UNUSUAL CASE REPORT

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ABSTRACT

Chicken pox is considered as benign infectious disease with variable ocular manifestations. We report a case of bilateral retinal vasculitis following chicken pox in a healthy 17-year-old male. The retinal manifestation is stabilized by treatment with oral acyclovir in combination with systemic steroids.

KEY WORDS

Acyclovir; chicken pox; steroids; vasculitis

INTRODUCTION

Ocular involvement after primary Varicella Zoster Virus (VZV) infection or chickenpox is rare and may involve any part of the eye from the conjunctiva to the optic nerve. Reported retinal vascular involvement in VZV infection include vasculitis, transient retinal arteriolitis, occlusive vasculopathy and recurrent multiple branch retinal artery occlusions.[1-3] We describe herein the case of bilateral ischaemic retinal vasculitis associated with chickenpox .

CASE DESCRIPTION

A healthy 17-year-old student from Begulsarai, India presented with a 2 week history of chickenpox in the out patient department . Ten days after the onset of cutaneous vesicular eruption, he experienced painless blurring of vision in his right eye. At the time of presentation, 3 days after the onset of symptoms, visual acuity in his right eye was 20/80. Examination of the anterior segment was unremarkable . Fundus examination revealed areas of pre-retinal and intra-retinal haemorrhages in the temporal and infero-temporal quadrant along with peri-venous sheathing and exudation [Figure 1 A]. Fluorescein angiogram of right eye revealed multiple areas of patchy capillary dropouts, diffused vascular leakage and staining [Figure 1 B] .

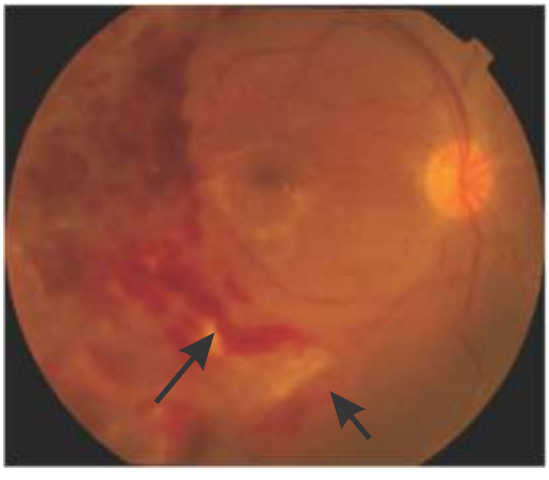

Figure 1A: Arrows showing peri-venous sheathing and exudation)

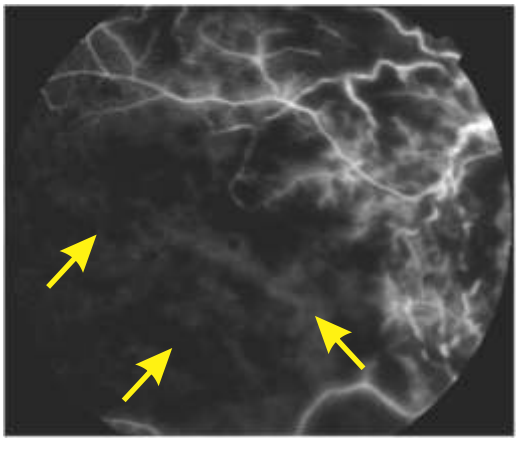

Figure 1B: (Arrows showing capillary dropout areas in fluorescein angiogram)

Visual acuity in his left eye was 20/20 with unremarkable anterior segment examination. Fundus evaluation of left eye revealed few areas of vascular sheathing in infero-temporal quadrant [Figure 2 A]. Fluorescein angiography of left eye revealed areas of peri-vascular leakage [Figure 2B]. The typical signs and symptoms of chickenpox had already subsided, except crusted eruptions on his face, limbs and abdomen that remained at the time of our examination [Figure 3A, Figure 3B]. He had no other relevant past history suggestive of immunocompromised status and had no high risk behaviours. Regarding his immunisation statu, we were unable to elicit a definite immunisation history but he had no personal or family history suggestive of any serious illness in the past. Both serum anti-VZV immunoglobulin M (5.88 Index Value) and immunoglobulin G (1689.0 mlu/ml) antibodies were elevated. VZV DNA was detected by polymerase chain reaction (PCR) from the vitreous cavity.

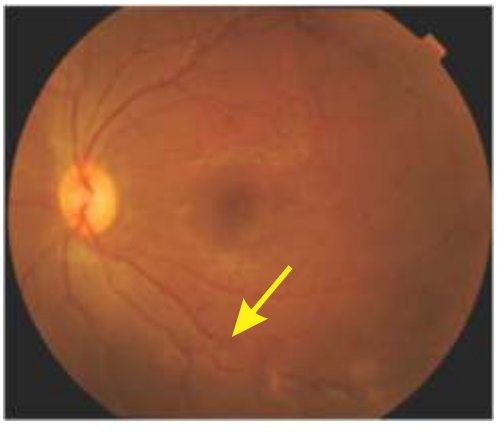

Figure 2A: (Infero-temporal quadrant showing vascular sheathing)

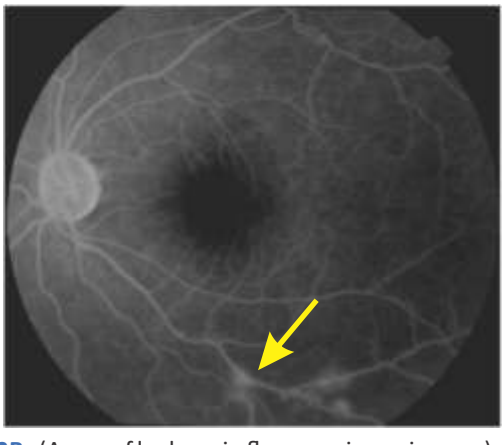

Figure 2B : (Areas of leakage in fluorescein angiogram)

Figure 3A

Figure 3B

Since the patient had not consulted any physician for his skin lesions and had not taken any anti-viral medication for his condition; there were features of active retinal involvement, a clinical diagnosis of chicken pox with retinal vasculitis was made and he was started on oral Acyclovir 800 mg five times per day induction phase for 2 weeks followed by maintenance phase 400 mg two times per day for 6 weeks along with oral steroid 1 mg / kg / day. At 2 weeks follow up, there was improvement in his visual acuity with improving vasculitis. However, the patient did not immediately follow up and he came back after 3 months with deterioration. At 3 months, his visual acuity RE was 20/200 with significant vitritis. Fundus evaluation of RE revealed active fibro-vascular tissue on the optic nerve head and along the vascular arcade with macular involvement. Rest of the anterior segment examinations, intra-ocular pressure, gonioscopy were unremarkable. He was re-started on oral Acyclovir along with oral steroid. Prophylactic laser was done over the non-perfused areas to prevent further ischaemia. Two weeks after re-starting of medication, the visual acuity RE improved to 20/125. There was significant improvement in vitritis and stabilization of fundus findings.

DISCUSSION

Chickenpox, caused by primary VZV infection, is infrequently associated with ocular inflammation and rarely with posterior segment inflammation. There are very few case series and case reports regarding the ocular manifestations of primary varicella infection. Gargouri et al[4] reported case series of five adults (seven eyes) with ocular involvement secondary to chickenpox. Anterior uveitis was the most frequently reported ocular manifestation. What is unique about our case, is that the patient had bilateral retinal vasculitis without anterior segment involvement which stabilized after the use of oral acyclovir. Although VZV is the most common causative agent of acute retinal necrosis (ARN) syndrome, this clinical entity has been rarely described following chickenpox infection. Matsuomo et al[1] reported case series of three patients with ARN following chicken pox. The clinical features in our case differed from ARN in the following points: (1) absence of anterior chamber and vitreous inflammation, (2) absence of classical necrotic retinal lesions.

There are only two case reports of retinal vasculitis associated with primary varicella infection. Poonyathalang et al[2] reported a case of unilateral ischaemic retinal vasculitis in a healthy adult following chickenpox. YH Kuo et al[3] reported a case of mild retinal vasculitis associated with primary chickenpox infection. Both cases had unilateral presentation unlike in our case which had bilateral involvement. Murdock et al[5] had reported a case of bilateral retinal vasculitis in an adult with chickenpox with systemic vasulitis. In our patient, increased varicella zoster virus IgM titre showed the immunological evidence of recent primary varicella infection which was further confirmed by positive PCR test of vitreous fluid .

It is uncertain whether this resulted from an immunologic response or the infectious process caused by VZV. No definite guideline for the management of such cases is available due to the limited number of cases reported. Treatment with systemic steroids alone or combined with antiviral agents has been described in previous case reports . Acyclovir has been used orally and intravenously in these patients. Other anti-virals used are gancyclovir, foscarnate.[6]

As these lesions resolve with or without anti-viral medications, the precise role of anti-viral is not clear. It is also unclear whether Zoster vaccinations prevent such clinical manifestation . There have been reports of patients developing Zoster ARN after Zoster vaccine administration.[7]

CONCLUSION

We report an immunocompetent 17 year old young adult who presented with bilateral retinal vasculitis after chickenpox. It is important to perform a detailed ocular examination in patients with varicella infection. Immediate recognition and proper treatment with systemic antiviral therapy may be essential for the prevention of severe sequelae and good visual outcome. However, a generalized comment cannot be made based on our single case. A large series is required to provide the correct guidelines for therapy.

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