Case report

Bilateral acute necrotizing retinitis due to cytomegalovirus infection in an infant: Challenging case report

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ABSTRACT

Purpose: To report a case of bilateral acute necrotising retinitis due to cytomegalovirus infection (CMV) in an Asian Indian infant.

Observations: An Asian Indian infant born with a birth weight of 1000 g at 26 week of gestation acquired cytomegalovirus infection from repeated blood transfusion for anemia. During the routine course of ROP screening, both eyes were detected with Type 1 ROP (stage 3 in zone 1 with plus disease) and treated with laser photoablation at 39 + 2 weeks post menstrual age. The disease responded to the laser and showed signs of regression. Four weeks after laser therapy (PMA 43 + 3 weeks), both eyes presented with vitritis, inferior vitreous condensation and white lesions in the lower nasal retina and temporal retina overlying the lasered retinal bed associated with white fluffy hemorrhagic lesions resembling necrotising retinitis. As the infant was seropositive for CMV earlier, Necrotising retinitis due to CMV was suspected. The CMV DNA was repeated and was detected in serum. Infant was treated with anti-CMV medication (oral valgancyclovir) for six weeks. The retinal lesions resolved completely.

Conclusion and Importance: Bilateral acute necrotising retinitis may present in an infant on post lasered retina as early as 16 weeks after birth. CMV may also present as acute necrotising retinitis in unusual cases therefore high index of suspicion and early management can result in a successful outcome.

1. Introduction

Acute necrotising retinitis presents as multifocal, well-demarcated areas of retinal necrosis of retina with rapid, circumferential progression of necrosis with occlusive vasculitis accompanied with vitritis. Varicella-zoster virus or herpes simplex virus type (HSV) 1 have been found to cause ARN in middle aged patients. Herpes simplex virus type 2 has been implicated to causes acute retinal necrosis in patients younger than 20 years. Acute retinal necrosis is a rare presenting feature of CMV retinitis. The mode of entry of CMV into endothelial and RPE cells in vivo has been demonstrated via endocytosis.

Retinitis with other systemic condition in a premature infant has been reported earlier. Co-existing ARN with ROP has been reported earlier but in both the cases the causative organism was herpes simplex virus 2. We are reporting a case of bilateral acute necrotising retinitisin an infant due to CMV infection.

2. Case report

A former 26 week gestational age, 1000 g preterm second born twin infant was evaluated at a tertiary level neonatal intensive care unit (NICU) on 34th day of life (PMA: 34 weeks) for ROP. On dilated fundus examination large avascular retina in zone 1 was present and infant was...
subsequently followed up. The infant had been previously treated for respiratory distress syndrome, patent ductus arteriosus and had undergone multiple blood transfusion for anemia of prematurity and had been discharged from NICU.

The infant had been previously being discharged from NICU after treatment for respiratory distress syndrome, patent ductus arteriosus and had underwent multiple blood transfusion for anemia of prematurity. The patient during the hospital stay had persistent anemia and thrombocytopenia for which blood transfusion was given. Persisting thrombocytopenia led to a suspicion of TORCH infection. The titres of serum CMV IgM (2.39 by CMIA) and IgG (151.65 au/ml by CMIA) were found to be reactive and CMV DNA (> 57.1 copies/ml) was detected in urine (real time PCR). The titres for toxoplasma, rubella, HSV 1 and HSV2 was negative. The baby was screened for CMV retinitis at PMA of 35 weeks but no lesion was detected. The infant was managed with supportive measures.
On follow up visit at PMA: 39 weeks bilateral ROP stage 3 plus disease in zone 1 was present along with popcorn lesions on dilated fundus examination (Fig. 1). The ROP was treated with fluorescein angiography (FA) guided laser.

The ROP along with popcorn lesions started to regress with time. After 1 month of laser treatment of ROP (PMA: 42 weeks), there was vitreous haze due to vitritis, inferior vitreous condensation and new white lesions in lower nasal periphery with white fluffy haemorrhagic patch (Figs. 2 and 3). In the light of these new findings the lesions were suspected to have peripheral retinal necrosis due to infective pathology. The fundus findings resembled necrotising retinitis. As the infant was seropositive for CMV earlier, necrotising retinitis due to CMV was suspected. The CMV DNA was repeated and was detected in serum. The dose of valganciclovir was tapered to 3.5 ml BD for 2 weeks. The CMV DNA was repeated and titres were found to be insignificant. Thereafter the treatment was stopped. On last follow up the patient had resolved lesions and vision was appropriate for age.

3. Discussion

Bilateral necrotising retinitis is a rare presenting feature of CMV. There has been report of coexistent ARN with ROP in a premature infant but CMV has not been implicated as an etiology so far.7

The infant was treated for ROP at PMA of 39 weeks for stage 3 plus disease in zone 1 according to standard protocol and avascular retinas was lasered. The development of vitritis, inferior vitreous condensation and new white lesions in lower nasal periphery with white fluffy haemorrhagic patch after 1 month of laser treatment could either be acute retinal necrosis, or necrotic changes secondary to laser or CMV. The absence of neovascularisation, recrudescence of existing vessel and existing vitritis were indicative of inflammation. The peripheral lasered retina developing CMV retinitis was intriguing as CMV has more predilection for retinal endothelial cells. A hypothesis regarding mechanism of CMV retinitis states that CMV gains entry into the retina through retinal pigment epithelial and endothelial cells by endocytosis. The presence of vitritis, inferior vitreous condensation, peripheral areas of retinal necrosis were features suggestive of ARN. Therefore, the diagnosis of necrotising retinitis due to CMV was made based on positive seropositivity and clinical picture.

Valganciclovir as therapy for CMV infection in an infant is debated. The first episode of CMV infection was managed conservatively. Oral valganciclovir was instituted in view of the acute necrotising retinitis and aggressive disease course. Vision could have been affected if treatment was delayed.

Early diagnosis and appropriate management resulted in resolution of retinitis. Therefore, a high degree of suspicion is needed to diagnose acute necrotising retinitis due to CMV in an infant.

Patient consent

A written informed consent was taken from patient’s legal guardian.

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Conflict of interest

The authors do not have any conflict of interest. All authors attest that they meet the current ICMJE criteria for Authorship.

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